Suid-Afrikaanse Tydskrif vir Geneeskunde : South African Medical Journal

Kaapstad 6 Februarie 1960 Deel 34 No. 6 Volume 34 Cape Town, 6 February 1960

SOME ASPECTS OF THE PATHOGENESIS AND SURGICAL MANAGEMENT OF PEPTIC ULCERS*

D. J. DU PLESSIS, CH.M., F.R.C.S., *Department of Surgery, University of the Witwatersrand, and Johannesburg General Hospital*

Surgical management depends on pathology, and in discussing the surgical approach to peptic ulcers we must base it on what is known of the pathogenesis of this condition. Peptic ulceration is not a single entity, but the common result of many disturbances, and successful management demands that we differentiate between the different types of peptic ulcers on the basis of the antecedent disorder.

The purpose of this paper is to discuss the different types of peptic ulcers with particular reference to their pathogenesis and to speculate about the value of the various methods of surgical treatment available for each one, with the emphasis on the risks of recurrent ulceration after the operation.

Peptic ulcers are produced by the corroding effect of acid and pepsin on the mucous membranes of different parts of the gastro-intestinal tract to which the acid and pepsin have access, and it is obvious that ulceration will occur as a result of either an excessive secretion of acid and pepsin, as occurs in duodenal ulceration, or a decrease in the resistance of the mucous membrane to an otherwise normal secretion of acid and pepsin, as is seen in gastric ulcers. In view of this, the logical form of surgical treatment for duodenal ulcers is a removal of the acid-producing area of the stomach[4] and operations for gastric ulcers should remove all the mucous membrane with a decreased resistance to acid and pepsin. In both types of operations care should be taken not to disturb the balance between acid secretion and mucosal resistance, because this will inevitably result in a recurrence of ulceration.

GASTRIC ULCERS

It is well known that the mucosal lining of the stomach can be divided into 3 different types—cardiac, fundic and pyloric. The fundic mucosa is encountered in the fundus and body of the stomach, whereas the cardiac region is found around the oesophageal opening and the pyloric region in the pyloric antrum. The pyloric and cardiac mucosae very closely resemble each other and for practical purposes can be described together.[5]

The fundic mucosa occupies the vast central area of the stomach, whereas the pyloric type of mucosa occupies on the average only 13·8% of the area of the adult stomach.[1] An interesting feature is that the pyloric mucosa extends further along the lesser curvature than along the greater curvature, and we thus find that in the adult the pyloric mucous membrane extends on the average 39·6% of the length of the lesser curvature, as opposed to only 13·2% of the length of the greater curvature[1] (Fig. 4). In absolute measures the pyloric area in the adult covers on an average 7·2 cm. of the lesser curvature and 5·2 cm. of the greater curvature.[1] It is possible, of course, that the pyloric mucous membrane might actually join up with the cardiac mucous membrane in some individuals, although I have not personally encountered such a case in human beings. Such a distribution of gastric mucous membrane is normal in certain animals like the horse and the pig, and consequently it is conceivable that it does occur in human beings in certain cases.

My interest in the aetiology of peptic ulcer commenced early in 1953, when during routine histological examination of gastric ulcers I was struck by the fact that none of these gastric ulcers occurred in an area with fundic mucous membrane and it thus seemed as if gastric ulceration was a disease of the pyloric mucous membrane only. This observation led me to examine 66 gastric ulcers microscopically and I found that in none of these cases was the mucous membrane adjoining the ulcers of the fundic variety. In all the cases there was a marked gastritis in the neighbourhood of the gastric ulcer, which frequently obscured the histological appearance to a certain degree. In 13 of these cases fundic mucosa was found very close to the ulcer, varying between 2 and 18 low-power fields away from the ulcer edge itself.

Fig. 1. A photomicrograph to illustrate chronic atrophic gastritis in pyloric mucosa.

* This article is based on papers presented at the 42nd South African Medical Congress (M.A.S.A.), East London, C.P., September-October 1959.

The reason for this susceptibility on the part of the pyloric mucosa to develop gastric ulceration seemed to be of fundamental importance, and I proceeded to investigate the matter further. Histological examination was made of 28 stomachs, resected for duodenal or gastric ulcers, by serial sections taken along the lesser and greater curves and the anterior and posterior walls. In each one chronic gastritis was encountered. The distribution of this gastritis was carefully plotted and the severity of the chronic gastritis was subdivided into chronic superficial gastritis and chronic atrophic gastritis.

Chronic superficial gastritis consists of an abnormal cellular infiltration with the formation of lymphoid follicles in the deeper layers of the mucosa. It may affect fundic and pyloric mucous membrane.

Chronic atrophic gastritis is a more advanced stage of gastritis[6] and results in flattening of the rugae and a granular mucosa which on microscopy, in addition to the features of superficial gastritis, shows varying grades of atrophy of the mucous membrane.

In the pyloric mucosa this atrophy results in extreme thinning of the mucous membrane to a stage where it is surprising that it can still resist ulceration (Fig. 1). In the fundic mucosa there is destruction of the chief and parietal cells, and the response to this irritation appears to be replacement of the damaged fundic glands by non-specific mucus-secreting cells arranged in coils to resemble the pyloric glands. These are called pseudo-pyloric glands.[2] This post-inflammatory metaplasia was clearly responsible for the original mistaken belief that gastric ulcers appeared only in the pyloric mucosa, because many gastric ulcers were found in fundic mucous membrane transformed by this pseudo-pyloric metaplasia (Fig. 2).

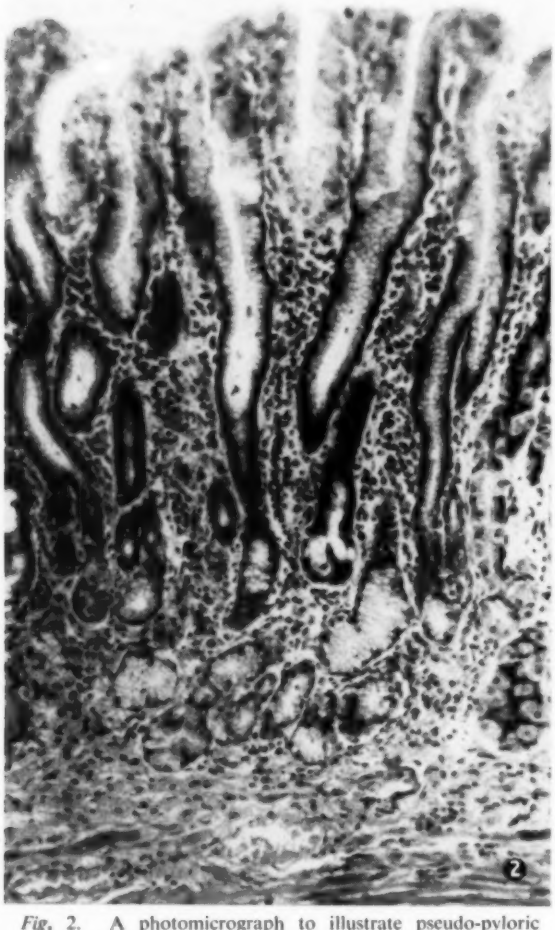


Fig. 2. A photomicrograph to illustrate pseudo-pyloric glands in fundic mucosa affected by chronic atrophic gastritis.

Another form of reaction is the formation of an intestinal type of mucosa[8,14]—intestinal metaplasia.

Sometimes the glands become distended to form small cysts. This may be due to destruction and subsequent repair with fibrosis and obstruction at the neck of the glands,[12] but at other times it is a result of gland regeneration following injury[31] (Fig. 3).

It was now obvious that the original concept was incorrect and that gastric ulcers may occur not only in pyloric mucosa but also in fundic mucosa which is the seat of chronic gastritis, and that the metaplasia described above disguises the fact that it is truly fundic mucosa. It is not surprising that such a mistake can be made if only a small portion of mucosa is available, because sometimes in severe atrophic gastritis it is impossible to decide whether one is dealing with pyloric or fundic mucous membrane.

Chronic gastritis is a very common if not invariable finding in stomachs resected for gastric ulceration[14] and the problem is to decide whether the gastritis is the primary event which leads to the development of gastric ulcer or whether it is the gastric ulcer which in turn produces the chronic gastritis in the surrounding gastric mucous membrane. Although it is impossible to give a dogmatic answer to this problem, there is nevertheless very suggestive evidence that the chronic gastritis is the primary event.[30,35] On many occasions I have encountered severe chronic gastritis of the superficial or the atrophic variety without any gastric ulcer present, whereas I have never encountered a gastric ulcer without severe surrounding chronic gastritis. The concept is thus that a chronic gastritis is the primary event and that a gastric ulcer may develop in this region.

It is now necessary to explain the cause of this chronic gastritis which is considered to be the precursor of the chronic ulcer. There is every indication that in many cases a chronic irritation of the surface of the gastric mucous membrane is certainly a very important factor. Such a chronic irritation can, of course, be caused by food taken by mouth, and a classical example of such a condition is chronic alcoholism,[6] where the recurrent irritation of strong liquor produces severe chronic gastritis. In this type of chronic gastritis the whole stomach is usually affected and often the fundic mucosa is more severely affected than the pyloric mucosa. Because of this extensive destruction of acid-producing cells, so little acid is secreted in the stomach that the formation of a chronic gastric ulcer is unlikely, and one thus not infrequently finds that these people do not present with a chronic gastric ulcer, but rather with diffuse atrophic gastritis and sometimes a superimposed acute gastritis (following a drinking bout) with resultant acute erosions and haemorrhage (Fig. 5).

Another known cause of chronic gastritis is gastric stasis, and this is the explanation for the fairly high incidence of

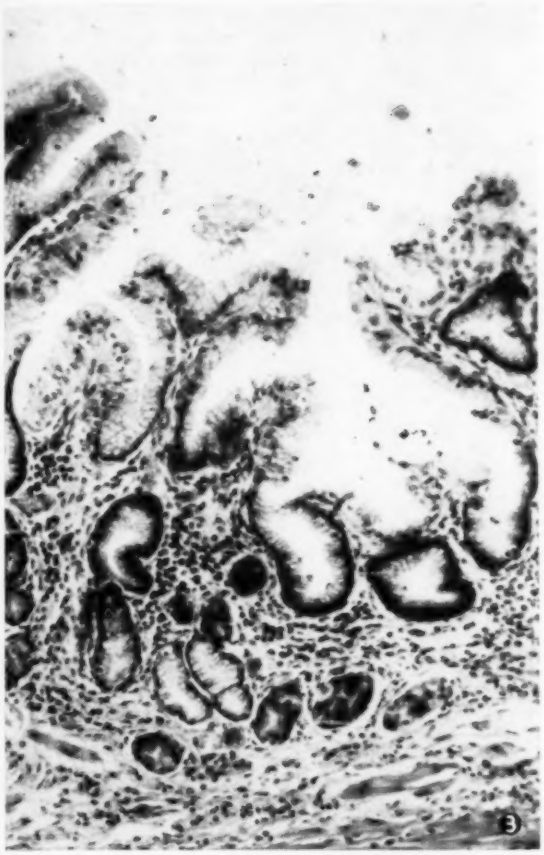


Fig. 3. A photomicrograph to show a microcyst in the gastric mucosa without any obstruction in the neck of the gland.

gastric ulcers after vagotomy without adequate drainage of the stomach.[9,10]

Another cause of chronic irritation to the mucous lining of the stomach occurs after gastrojejunostomy where there is repeated reflux of bile and pancreatic juice into the stomach.[12,13] This persistent irritation produces a chronic gastritis which sometimes goes on to produce a chronic gastric ulcer or haematemesis from multiple superficial erosions. The gastritis produced by a gastrojejunostomy is centred around the stoma, and there seems very little doubt that it has been produced by the reflux of bile and pancreatic secretion through the stoma (Fig. 6).

In other cases of gastric ulcer one finds that the chronic gastritis extends from the pylorus proximally for a variable distance into the stomach, usually confined to the pyloric antrum and lowermost portion of the fundus[14,30] and mostly along the lesser curvature[2,3] (Fig. 7). It is a common condition, increasing in extent and severity with increasing age.[30] The remainder of the gastric mucous membrane appears normal. It is thus suggestive that the factor producing the chronic irritation arises in the region of the pylorus and extends for a variable distance proximally. It seems thus highly unlikely that this form of irritation has been produced by food or fluid taken by mouth and, in view of the fact that it seems likely that reflux of bile and pancreatic secretion through a gastrojejunostomy stoma can produce surrounding chronic gastritis, it is suggested that this type of chronic gastritis, extending from the pylorus proximally, has arisen as a result of the reflux of bile and pancreatic secretion through an incompetent pylorus.

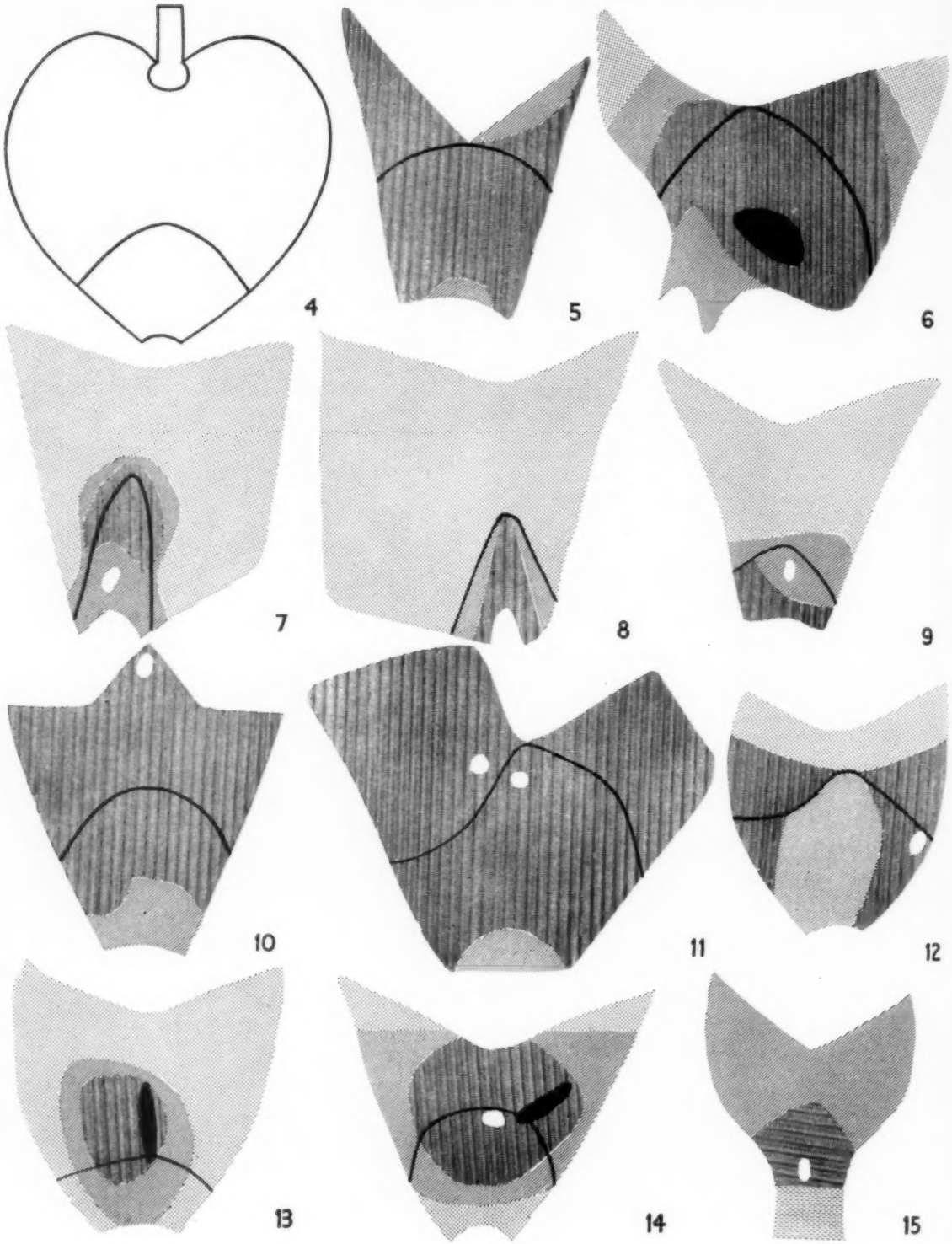
It has previously been pointed out that the pylorus is not a sphincter which prevents gastric emptying, but that its purpose is to prevent the reflux of bile and pancreatic secretion back into the stomach when duodenal contraction takes place.[7] If this function is inadequate then bile and pancreatic secretion can reflux into the stomach during duodenal contraction, in which case the distal portion of the stomach will be constantly irritated by this fluid. It is thus suggested that this type of gastric ulcer is the result of an incompetent pyloric sphincter which allows duodenal reflux and in that way produces a chronic gastritis in the distal portion of the stomach, which is the precursor of the chronic gastric ulcer in that region.

That such a condition probably exists in many people is shown in a review of a large number of cases in which it was found that only 27·8% of stomachs of normal people showed absence of gastritis,[3] and one has found it in the presence of duodenal ulcers[8] (Fig. 8). This gastritis always extends from the pylorus proximally and it is maximal along the lesser curvature, presumably because the tone of the stomach opposes the anterior and posterior stomach walls, leaving only a small region along the lesser curvature for fluid to reflux, in exactly the same way as liquid taken by mouth runs down along the lesser curvature. The presence of such a chronic gastritis in the pyloric region of the stomach largely centred on the lesser curvature is a common finding, and is not an indication of a diffuse gastritis of the stomach.[8]

If it is accepted that this type of irritation is a common feature then, of course, it is easy to understand how in selected cases a chronic gastric ulcer will occur in the pyloric region of the stomach, particularly along the lesser curvature. It is also easy to understand that in some cases a duodenal ulcer occurs in the presence of a gastric ulcer[11] because there is nothing to prevent a hypersecreting stomach from also having an incompetent pyloric sphincter, thus allowing sufficient reflux to produce a chronic gastritis along the lesser curvature with the resultant formation of a chronic gastric ulcer. In these cases the extent of the chronic gastritis is limited; otherwise there would not be enough secretory cell mass to produce sufficient gastric secretion to cause a duodenal ulcer (Fig. 9).

An interesting speculation now is to decide why gastric ulcers particularly occur in the region of the pyloric mucosa. Is it merely because the pyloric mucosa is closer to the pylorus and that it is thus likely to be most severely affected by the refluxing duodenal contents, or is the pyloric mucosa in fact less resistant to this refluxing fluid so that a gastric ulcer is produced in this area in preference to the fundic mucosa? There has been some suggestion that the pyloric mucosa close to its junction with the fundic mucosa is especially susceptible to ulceration,[32,34] and this will explain the classical location of gastric ulcers, but it is by no means invariable, and we know that a gastric ulcer may be found more proximally in fundic mucosa[30,32,33] (Fig. 10).

In 3 cases studied with severe atrophic gastritis affecting the pyloric and fundic mucosa, a gastric ulcer was found in


4
5
6
7
8
9
10
11
12
13
14
15

the pyloric region in 2 cases and in the fundic mucosa in 1. On the other hand, I have found a person with two gastric ulcers, one in the pyloric and one in the fundic region (Fig. 11).

One interesting finding has been to see the junction between fundic and pyloric mucous membranes and to find that the body mucosa shows superficial gastritis only and the pyloric mucosa immediately adjoining it shows severe atrophic gastritis, suggesting that the latter area is more susceptible to irritation (Fig. 17). When performing a gastric resection for a gastric ulcer, therefore, it is important that the surgeon should remove an extensive area of the lesser curvature so as to remove the pyloric mucous membrane and the area which has been subjected to gastritis quite irrespective of the situation of the gastric ulcer. For this reason I recommend that in all gastrectomies performed for gastric ulceration a Pauchet manœuvre should be performed to ensure that all the susceptible and damaged mucous membrane is removed at the time of the operation to prevent the recurrence of another gastric ulcer. If this is not done then damaged mucosa is left behind which may produce another gastric ulcer.

In these cases the duodenum is normal and therefore there is no increased chance of a duodenal ulcer occurring later. There is thus no particular need to perform a Polya type of operation and I consequently prefer a Billroth I operation because it eliminates the afferent loop, which is the source of potential trouble at a later stage.[7] There is of course the possibility of duodenal reflux into the gastric remnant with the formation of a new gastric ulcer, and for that reason I wonder whether a Roux-Y type of operation is not preferable in this type of lesion.

During this investigation an interesting observation was encountered in connection with the effect of cortisone on the development of peptic ulcers. It is known that cortisone increases the gastric secretion and in such an event will be likely to produce a duodenal ulcer if the gastric mucosa is normal.[4,15] If, however, cortisone is administered to a person with an incompetent pyloric sphincter which allows free reflux and the development of chronic gastritis, the anti-inflammatory effect of cortisone will result in severe gastric mucosal atrophy with very little inflammatory reaction; one such case was encountered where a marked feature was the gross chronic atrophy of the gastric mucosa with virtually no inflammatory reaction (Fig. 16). This patient actually developed a spontaneous gastrocolic fistula and no doubt this extension of gastric ulcer into the colon was also the result of this lack of inflammatory reaction to the irritation produced by the duodenal reflux (Fig. 12). It thus seems that the administration of cortisone may be responsible for the development of either a duodenal ulcer or a gastric ulcer, depending on the function and efficiency of the stomach and the pyloric sphincter.

Figs. 4-15.

Stippled area	= normal gastric mucous membrane
Interrupted transverse lines	= normal duodenal mucous membrane
Diagonal lines	= chronic superficial gastritis
Wavy lines	= chronic atrophic gastritis
Black areas	= gastro-enterostomy stoma
White areas	= gastric ulcers

Thick black line outlines the junction between pyloric and fundic mucosa. These diagrams illustrate the following:

Fig. 4. The average distribution of cardiac, fundic and pyloric mucosa in an adult stomach.
Fig. 5. The distribution of gastritis in a stomach resected for haematemesis in a European male aged 39 who was a chronic alcoholic. (No chronic ulcer was present.)
Fig. 6. The distribution of gastritis in a stomach resected for haematemesis in a Coloured male aged 43 who had had a previous gastro-jejunostomy. (No chronic ulcer was present.)
Fig. 7. The distribution of gastritis in a stomach resected for a chronic gastric ulcer. (Coloured male aged 49.)
Fig. 8. The distribution of gastritis in a stomach resected for a chronic duodenal ulcer. (European male aged 43.)
Fig. 9. The distribution of gastritis in a stomach resected for active chronic gastric and duodenal ulcers. (Coloured female aged 46.)
Fig. 10. The distribution of gastritis in a stomach resected for a chronic gastric ulcer high up on the lesser curvature. (European male aged 68.)
Fig. 11. The distribution of gastritis in a stomach resected for two chronic gastric ulcers. (European male aged 42.)
Fig. 12. The distribution of gastritis in a stomach resected for a chronic gastric ulcer and gastrocolic fistula in a European female aged 41 who had received large doses of cortisone for the previous 7 years.
Fig. 13. The distribution of gastritis in a stomach resected for a jejunal ulcer following a previous gastro-enterostomy in a European male aged 69.
Fig. 14. The distribution of gastritis in a stomach resected for a gastric ulcer following a previous gastro-enterostomy in a European male aged 62.
Fig. 15. The distribution of gastritis in a stomach resected for a gastric ulcer following a previous Billroth I gastrectomy for a duodenal ulcer in a European male aged 51.

DUODENAL ULCER

This condition is now usually considered to be due to excessive secretion of acid and pepsin by a stomach which contains an abnormally large number of acid-secreting cells.[16,17]

The increased volume of acid and pepsin produced by this increased parietal-cell mass will, of course, pass over the pyloric mucous membrane of the stomach and then down the bowel, and the fact that it produces an ulcer only in the first part of the duodenum indicates a relatively poor resistance in that area.

The defence of the fundic mucous membrane of the stomach against this high acid secretion is inherent in the mucosa and it is clearly very high because ulceration never occurs in fundic mucosa which is still intact. The pyloric mucosa is protected by the mucus and alkali secreted by the pyloric glands, and this too is a very efficient protective mechanism, for it is unusual to find a gastric ulcer in the presence of an abnormally high acid secretion. Sometimes this does happen if there is a localized gastritis which lowers the resistance of that area without interfering with the acid-secreting mechanism. Under these circumstances we then find the comparatively rare combination of an active duodenal ulcer and an active gastric ulcer present at the same time.

Beyond the ampulla of Vater there are virtually no Brunner's glands, and the protection of the bowel beyond that point against acid and pepsin depends entirely on the neutralizing and buffering effect of the bile and pancreatic secretion.

The first portion of the duodenum up to the ampulla of Vater depends for its protection on the secretion of mucus and alkali by the Brunner's glands, which closely resemble the pyloric glands histologically,[5] and duodenal ulceration may thus be considered to be due to an imbalance between the acid-secreting cells of the stomach and the Brunner's glands. It is known that a congenital inadequacy of the Brunner's glands may occur[5] and this may explain those cases of duodenal ulceration without an abnormally high

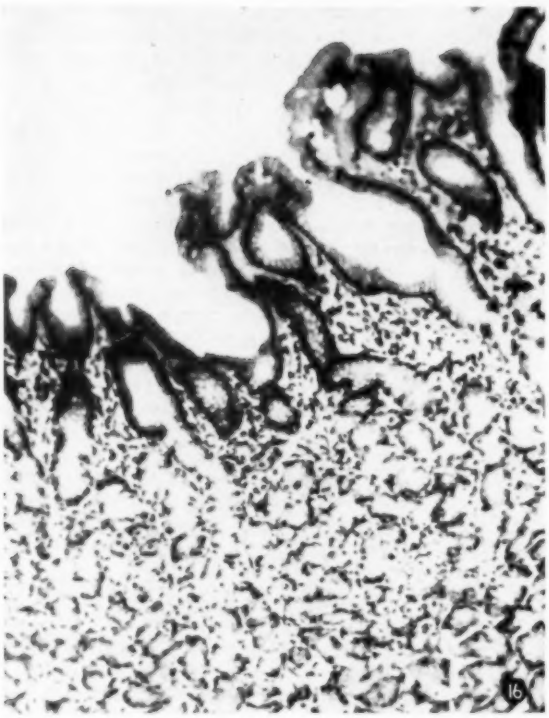


Fig. 16. A photomicrograph showing atrophy of the fundic mucosa without significant cellular infiltration in a European female aged 41 with a gastrocolic fistula. She had been on large doses of cortisone for 7 years.

acid secretion.[34] In addition, it should be kept in mind that there is a severe duodenitis present in cases with duodenal ulceration, and mucosal thinning and atrophy of the Brunner's glands[35] (Fig. 18). Whether this is the cause or the effect of the duodenal ulcer is uncertain, but the end result in any event would be a lowering of the mucosal resistance, and consequently any operation which does not divert acid and pepsin away from the duodenum will be liable to be followed by a recurrence of a duodenal ulcer. This is probably the explanation for the comparatively high incidence of duodenal ulceration after a Billroth I operation performed for duodenal ulceration[18] and for the disappointing results with sleeve resections, which at first glance would appear to be the most logical operation for a duodenal ulcer.[19-29] For the present it thus seems as if the Polya type of gastrectomy is the most reliable operation for duodenal ulceration, although this type of operation results in the reflux of bile and pancreatic secretion into the gastric remnant, which may develop a gastritis and in time a gastric ulcer. It seems likely that a Hoffmeister valve will assist in directing the bile away from the gastric remnant into the efferent loop and consequently I feel that this refinement should be used when performing a Polya type gastrectomy.

STOMAL ULCER

Just as a duodenal ulcer is produced by a hypersecreting stomach which overcomes the resistance of the duodenal mucosa, and many gastric ulcers by reflux of bile and pancreatic juice which produces chronic gastritis, so we can also subdivide stomal ulceration after gastrojejunostomy or gastrectomy into two varieties. The one variety will be produced by a hypersecreting stomach which produces irritation of the jejunum leading to a jejunal ulcer (Fig. 13). On the other hand the stoma allows free reflux of pancreatic secretion into the stomach, producing the surrounding chronic gastritis which will be the precursor of a stomal ulcer on the gastric side of the suture line (Fig. 14). This, of course, may occur after a Billroth I operation too (Fig. 15).

The differentiation between these two types of stomal ulcers is important because the management is naturally quite different.

If the stomal ulcer is in the jejunum then it is obvious that an excessively high volume of acid and pepsin is being secreted by the stomach, and treatment should be directed at removing this high volume of acid from the susceptible jejunum. If the stomal ulcer is on the gastric side of the suture line, then clearly the treatment should not be directed at decreasing acid secretion but at the removal of the affected gastric mucous membrane and a prevention of a repetition of the same occurrence by diverting bile and pancreatic secretion away from the stomach, as with a Roux-Y type of anastomosis.

What has been said is obviously highly theoretical and dependent on certain experimental data which are as yet unproved. More proof is obviously necessary before these factors can be established as fact, and therefore I hope that this paper will be looked upon as a preliminary and provisional report. We are still in the stage of observation as far as the correct surgical treatment of peptic ulcers is concerned and we are not yet in a position to formulate a definite hypothesis. We must thus be careful when attempting to do so, because a misconception may impede the progress of knowledge. Any hypothesis must be subjected to repeated and stringent criticism and it is in that spirit that I present this paper.

SUMMARY

1. Gastric ulcers occur in mucous membrane damaged by chronic gastritis.

2. There are many causes of chronic gastritis, but a striking type is seen around the stoma of a gastro-enterostomy, presumably caused by the irritation of refluxing bile and pancreatic secretion.

3. Many cases have been found with chronic gastritis in the distal part of the stomach extending from the pylorus for a variable distance proximally. It is suggested that this may be due to reflux of bile and pancreatic secretion through an incompetent pyloric sphincter.

4. The majority of gastric ulcers are found in pyloric mucosa. This may be due to the distal situation of this type of mucosa, but there is some evidence that pyloric mucosa is less resistant to irritation than fundic mucosa.

5. Not all gastric ulcers occur in pyloric mucosa. Some are obviously in fundic mucosa, whereas others are in fact in fundic mucosa but the pyloric metaplasia of chronic gastritis disguises this fact.

6. Surgical treatment of gastric ulcers must include removal of all affected mucous membrane.

7. In cases with duodenal ulceration there is a congenital or acquired defect in the duodenum and for that reason a

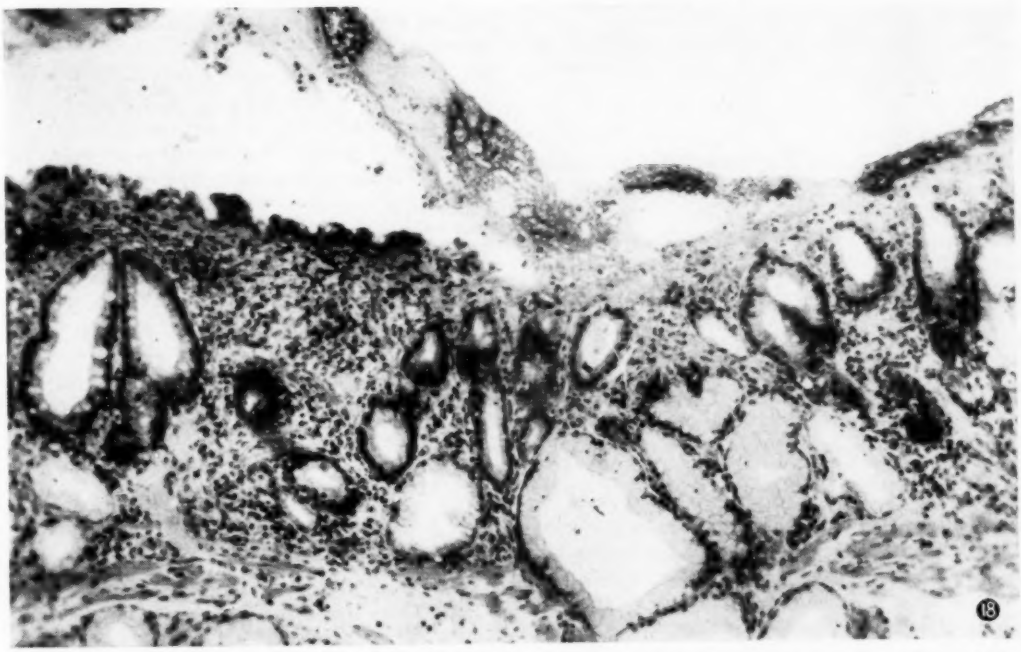


Fig. 17. A photomicrograph to illustrate an apparent increased susceptibility of the pyloric mucosa to gastritis. The pyloric mucosa is very thin and atrophic whereas the adjoining fundic mucosa shows only superficial gastritis.
Fig. 18. A photomicrograph showing severe duodenitis in a patient with a chronic duodenal ulcer.

Billroth I gastrectomy or a tubular resection will result in a recurrence of a duodenal ulcer in many cases.

8. A stomal ulcer may be in the jejunum, due to excessive acid secretion, or on the gastric side of the stoma following on the chronic gastritis produced by the refluxing bile and pancreatic secretion. The surgical treatment will vary according to the site of the stomal ulcer.

REFERENCES

1. Landboe-Christensen, E. (1944): Acta path. microbiol. scand., suppl. **54**, 671.
2. Wood, I. J. and Taft, L. I. (1958): *Diffuse Lesions of the Stomach.* London: Arnold.
3. Hebbel, R. (1949): Amer. J. Path., **25**, 125.
4. Bruce, J., Card, W. I., Marks, I. N. and Sircus, W. (1959): J. Roy. Coll. Surg. Edin., **4**, 85.
5. Landboe-Christensen, E. (1944): Acta path. microbiol. scand., suppl. **52**.
6. Doig, R. K. and Wood, I. J. (1952): Med. J. Austral., **1**, 593.
7. du Plessis, D. J. (1956): Med. Proc., **2**, 29 and 77.

8. Morson, B. C. (1955): Lancet, 2, 1068.
9. Dragstedt, L. R. *et al.* (1955): Arch. Surg., 71, 136.
10. Oberhelman, H. A. and Dragstedt, L. R. (1955): Surg. Gynec. Obstet., 101, 194.
11. Johnson, H. D. (1955): Lancet, 1, 266.
12. Palmer, E. D. (1954): Medicine, 33, 199.
13. *Idem* (1948): Gastroenterology, 10, 671.
14. Magnus, H. A. (1937): J. Path. Bact., 44, 389.
15. Dragstedt, L. R. (1956): Gastroenterology, 30, 208.
16. Hunt, J. N. and Kay, A. W. (1954): Brit. Med. J., 2, 1444.
17. Cox, A. J. (1952): Arch. Path., 54, 407.
18. Walters, W. (1957): Arch. Surg., 74, 615.
19. Tanner, N. C. (1954): Postgrad. Med. J., 30, 523.
20. Kay, D. W. (1956): J. Roy. Coll. Surg. Edin., 2, 54.
21. Woodward, E. R. *et al.* (1956): Arch. Surg., 72, 1003.
22. Connell, F. G. (1929): Surg. Gynec. Obstet., 49, 696.
23. Kaplan, R. S. and State, D. (1956): Amer. J. Gastroent., 26, 29.
24. Wangensteen, O. H. (1956): *Ibid.*, 26, 37.
25. Berne, C. J. (1954): *Ibid.*, 22, 126.
26. Thal, A. P., Perry, J. F. and Wangensteen, O. H. (1957): Surgery, 41, 576.
27. Wangensteen, O. H. (1957): *Ibid.*, 41, 686.
28. Zollinger, R. M. and Ellison, E. H. (1955): Ann. Surg., 142, 709.
29. Gray, S. J., Ramsey, C. G. and Reifenstein, R. W. (1954): New Engl. J. Med., 251, 835.
30. Stout, A. P. (1947): Bull. N.Y. Acad. Med., 23, 101.
31. *Idem* (1945): N.Y. St. J. Med., 45, 973.
32. Oi, M., Oshida, K. and Sugimura, S. (1959): Gastroenterology, 36, 45.
33. Karsner, H. T. (1949): *Human Pathology*. Philadelphia: J. B. Lippincott.
34. Marks, I. N. and Shay, H. (1959): Lancet, 1, 1107.
35. Collins, V. P. (1943): Ann. Surg., 118, 1005.

A NOTE ON PRIMING THE DEWALL OXYGENATOR

MALCOLM B. MCKENZIE, M.B., CH.B. (CAPE TOWN), *Registrar, Department of Surgery, University of Cape Town and Red Cross War Memorial Children's Hospital*, and
CHRISTIAAN N. BARNARD, M.D., M.MED. (CAPE TOWN), M.S., PH.D. (MINNESOTA), *Senior Clinical Lecturer, University of Cape Town; Senior Surgeon, Groote Schuur Hospital; Director of Surgical Research, Department of Surgery, University of Cape Town*

In a previous communication we described in detail the assembly and priming of the helix-reservoir bubble oxygenator.[1] The methods used, though satisfactory in their end results, are time-consuming. Since the report of Faggella *et al.*,[2] however, describing the use of saline at 90°C, and with our own further experience with this oxygenator, we have reduced the time of this operation to a few minutes.

METHOD

(*a*) *Initial Priming with Saline*

A vacolitre of normal saline is placed in a boiling sterilizer for 30 minutes, and the helix, blood filter and arterial line are assembled as we previously described.[1] The vacolitre is then removed from the sterilizer and its contents, which we have found to have a temperature of approximately 90°C at this stage, are poured into the top end of the helix.

The hot saline flows through the helix and filter and, once the latter has been filled, a clamp is applied to the arterial line distal to the filter. Filling is then continued until the helix contains approximately 800 ml. of fluid, when a second clamp is applied to the arterial line proximal to the filter, and the saline in the system is inspected for bubbles.

It is found that few bubbles form when hot saline is employed for priming, and any bubbles present are easily dislodged by tapping the helix and filter with a patella hammer. Once the helix and filter have been cleared of bubbles, the clamps on the arterial line are removed and the saline is allowed to flow slowly through the remainder of the system. The metal connectors and opaque section of latex tubing in the circuit are beaten to dislodge any bubbles which may have been trapped, and a clamp is applied to the distal end of the arterial line.

If the temperature of the priming fluid is below 90°C, bubble formation is increased and, if the temperature of this fluid is allowed to fall appreciably before de-bubbling is commenced, it is found that bubbles are more difficult to dislodge. It is therefore important to perform the priming as swiftly as possible, and to complete the whole operation in a few minutes.

Calibration of the arterial pump and pump occlusion are carried out as described before.[1]

(*b*) *Priming with Compatible Donor Blood*

As soon as the pericardial sac has been opened and the necessity for direct-vision surgery confirmed by the surgeon, priming is commenced. Donor blood is pumped from a blood bottle, by means of the venous pump, into the mixing chamber, where it is mixed with 100% oxygen. The venous pump is adjusted so that it is only just propelling the blood forward. The oxygen flow through the mixing chamber is regulated so that the oxygen bubbles are seen only just rising in this chamber. The blood-oxgyen mixture therefore enters the de-bubbling chamber, where the bubbles are broken down very slowly, and a stream of blood trickles gently into the helix, causing no turbulence, and thus no bubbles form on the blood surface within the helix.

Once the helix contains approximately 800 ml. of blood, free of bubbles, both the venous pump and the oxygen flow rate are stepped up, and the remainder of the priming volume of blood is pushed through the system more rapidly. Should bubbles form now, they are of no significance, and will tend to gravitate upwards when perfusion is commenced.

CONCLUSION

Priming the oxygenator with normal saline at 90°C takes but a few minutes, does not affect the transparency of the Mayon tubing, and appears to be entirely satisfactory.

Priming the oxygenator with blood as described prevents bubble formation within the helix, and hence prevents consequent unforeseen delay during surgery.

SUMMARY

The problem of bubble formation during the operation of priming the helix-reservoir bubble oxygenator is discussed. A method of obviating this complication is described.

We wish to thank Mr. C. C. Goosen, Mrs. V. M. Connell, and Miss D. M. Shepstone, for their assistance in perfecting the method of priming described.

REFERENCES

1. McKenzie, M. B. and Barnard, C. N. (1958): S. Afr. Med. J., 32, 1145.
2. Faggella, M. L., Zuhdi, M. N., Greer, A. E., Carey, J. M. and Schilling, J. A. (1958): Amer. J. Surg., 96, 696.

Suid-Afrikaanse Tydskrif vir Geneeskunde : South African Medical Journal

VAN DIE REDAKSIE : EDITORIAL

VERNOUING VAN DIE ARTERIA RENALIS

In die meerderheid van gevalle van verhoogde arteriële bloeddruk is die oorsaak en patogenese onbekend en word daar met die benaming ‚selfstandige' of ‚essensiële' hipertensie volstaan. Ten spyte van groot vooruitgang by die behandeling van hierdie groep gevalle, bly die behandeling palliatief en nie genesend nie. Onder die bekende oorsake van arteriële hipertensie is glomerulonefritis belangrik, maar ongeneeslik. In 'n betreklike klein minderheid van gevalle kan 'n bepaalde oorsaak, wat wel soms geneesbaar is, vir die arteriële hipertensie gevind word, bv. koarktasie van die aorta (hipertensie in boonste deel van die liggaam), feokromositoom, primêre aldosteronisme, Cushing se sindroom, en sekere gevalle van chroniese atrofiese piëlonefritis of hipoplasie van die nier. By hierdie klein reeks oorsake wat soms geneesbaar is, is in die afgelope jare bygevoeg vernouing van die arteria renalis of een van sy takke.[1]

Goldblatt en sy medewerkers[2] het reeds in 1934 die aandag gevestig op verhoogde arteriële bloeddruk wat in eksperimentele diere veroorsaak kon word deur die arteria renalis met 'n klem te vernou. Ten spyte van 'n groot hoeveelheid navorsingswerk op hierdie gebied is die Goldblatt-meganisme nie algemeen aanvaar as die oorsaak van ‚essensiële' hipertensie by die mens nie. Tog is daar in die afgelope jare verskeie gevalle beskryf waarin vernouing van die arteria renalis verhoogde arteriële bloeddruk verooksaak het, net soos die vernouende klem van Goldblatt by sy diere. Hoewel seldsaam, is hierdie gevalle in dié sin belangrik dat hulle soms volkome genees kan word.

Die belangrikste oorsake van vernouing van die arteria renalis is aangebore afwykings en aterosklerose. In albei gevalle kan die arteria renalis, sy uitmonding in die aorta, of die aorta self aangetas word. Soms is daar 'n gepaardgaande aneurisma van die arteria renalis. Trombose, embolisme of selfs besering mag ook 'n rol speel. Solank daar vernouing veroorsaak word, kan ernstige arteriële hipertensie ontstaan ongeag die patologiese aard van die oorsaak van die vernouing.

Die simptome en tekens is die wat in die geval van enige ernstige hipertensie gevind word. Die sistoliese sowel as die diastoliese bloeddruk is aansienlik verhoog. Af en toe word geruise oor die aorta of arteriae femorales gehoor. Die gewone chemiese en mikroskopiese ondersoeke van die urine toon meestal geen afwyking nie. Die bloedureum en ureum-opruimingstoets is ook gewoonlik binne normale perke. Röntgenondersoeke van die niere self, insluitende piëlografie, toon dikwels 'n heeltemal normale beeld, behalwe dat die twee niere soms nie ewe groot is nie. Nog meer gespesialiseerde ondersoeke is nodig om die fout aan te dui, naamlik:

(1) Differensiële nier-opruimingstoetse, bv. met inulin of para-aminohippuursuur, mag 'n verskil tussen die twee niere aandui, waarvan daar afgelei kan word watter een aangetas is.

(2) Renale arteriografie, deur aortagrafie (translumbaal of opwaarts vanuit die arteria femoralis—Seldinger), toon gewoonlik die vernouing van die arteria renalis. Dit is op die oomblik die akkuraatste diagnostiese ondersoekmetode.

Dit is dus duidelik dat dit moeilik is om in 'n bepaalde geval met arteriële hipertensie te bewys dat vernouing van die arteria renalis die oorsaak van die toestand is, behalwe deur middel van aortagrafie. Hoewel aortagrafie veel minder gevaarlik is as wat die deursnee geneesheer dink, is dit nie heeltemal sonder risiko nie en sou dit nie op die oomblik aanvaar word as 'n geregverdigde roetineondersoek vir alle gevalle van hipertensie nie. Om te besluit watter gevalle aortagrafies ondersoek moet word, is moeilik, maar die volgende kan as leidraad dien, omdat (nadat ander oorsake uitgesluit is) hulle in die rigting van vernouing van die arteria renalis as oorsaak wys:

(i) Die verskyning van hipertensie voor dertigjarige ouderdom (aangebore afwykings).

(ii) Die verskyning van hipertensie vir die eerste maal na vyf-en-vyftigjarige ouderdom (aterosklerose).

(iii) Die skielike verergering van bestaande hipertensie.

(iv) Waar 'n sistoliese arteriële geruis in die buik hoorbaar is.

(v) Waar 'n verskil in die grootte van die twee niere nie aan ander oorsake toegeskryf kan word nie.

Chirurgiese behandeling is dikwels moontlik mits die hipertensie nie reeds so lank teenwoordig was dat onomkeerbare veranderinge veroorsaak is nie. In hierdie verband is dit interessant dat by eensydige vernouings die nier met die normale arteria renalis gewoonlik erger onder die hipertensie ly as die een met die vernouing, omdat laasgenoemde deur die vernouing teen 'n te hoë druk beskerm word. Dit is 'n besondere rede waarom daar probeer moet word om die arteriële lumen te herstel en die nier te behou liewer as om dit net te verwyder. Chirurgiese ingrepe op die arteria renalis sluit in: Eksisie van stenose en hegting; lienorenale arteriële anastomose; aortarenale transplantasie, en endarterektomie. Waar vatherstel nie moontlik is nie, en die toestand van die ander nier bevredigend is, word nefrektomie aangedui. By geskikte gevalle is die resultate dramaties en kan van *genesing* van die toestand gepraat word; by minder geskikte gevalle word *opvallende verbetering* dikwels behaal.

Hoewel hierdie toestand op die oomblik selde gevind word, is dit moontlik dat fynere chemiese en röntgenologiese ondersoeke mettertyd meer sulke gevalle aan die lig sal bring. Daar is trouens reeds spekulasie oor die moontlikheid dat hierdie meganisme in takke van die arteria renalis en selfs in die klein intrarenale vate vir ‚essensiële' hipertensie verantwoordelik kan wees. Vir hierdie teorie ontbreek die bewys op die oomblik, maar vir die enkele pasiënte

by wie die bloeddrukverhoging deur vernouing van die nierslagaar veroorsaak word, is dit van groot belang om die toestand te herken en chirurgies te behandel.

1. DeCamp, P. T., Birchall, R. en Batson, H. (1958): Wetenskaplike Uitstalling by 'n vergadering van die Amerikaanse Mediese Vereniging in San Francisco.
2. Goldblatt, H., Lynch, J., Hanzal, R. F. en Summerville, W. W. (1934): J. Exper. Med., 59, 374.

NARROWING OF THE RENAL ARTERY

In most cases of raised arterial blood pressure the aetiology and pathogenesis of the condition are obscure and the label 'essential hypertension' is used. Moreover, treatment in cases of essential hypertension is still often palliative and not curative, in spite of great progress in this field. Glomerulonephritis is one of the known causes of arterial hypertension, but it is incurable. In a minority of cases a definite cause for the hypertension can be found, viz. coarctation of the aorta, phæochromocytoma, primary aldosteronism, Cushing's disease, and in some cases chronic atrophic pyelonephritis or hypoplasia of the kidney. In some of these cases the hypertension can be cured.[1]

During recent years the condition of narrowing of the renal artery has been added to the short list of curable hypertensive conditions. Goldblatt and his associates[2] have already (1934) drawn attention to the raised arterial blood pressure which could be brought about in experimental animals by clamping the renal artery. This mechanism, suggested by Goldblatt, has not been generally accepted as a cause of 'essential hypertension' in Man, in spite of a great deal of research work in this field. Yet, a number of cases have been described during the past years in which narrowing of the renal artery caused raised arterial blood pressure—like the clamp in Goldblatt's animals. These cases, although rarely encountered, are nevertheless important because they can often be cured completely.

Congenital malformations and atherosclerosis are some of the most important causes of narrowing of the renal artery. In both cases the renal artery can be involved, or its junction to the aorta, or the aorta itself. An associated aneurysm of the renal artery is sometimes present. Thrombosis, embolism and even injury may also be causative factors. A serious degree of arterial hypertension can arise while narrowing of the artery is present, irrespective of the pathological nature of the narrowing.

The symptoms and signs are those found in any severe hypertensive condition. Both the systolic and diastolic blood pressures are raised considerably. Murmurs are sometimes heard over the aorta or the femoral arteries. The usual chemical and microscopical examinations of the urine are generally normal. The blood urea and the urea-clearance test are also, as a rule, within normal limits. X-rays of the kidneys, including pyelography, often fail to show any abnormality except perhaps an inequality of the two kidneys. The following special investigations are necessary to make the diagnosis, viz.

1. Differential clearance tests, e.g. inulin or para-aminohippuric acid may indicate a difference between the two kidneys—from which a deduction can be made regarding which artery is affected.

2. Renal arteriography (through a translumbar route or through the aorta or upwards from the femoral artery—Seldinger), usually shows narrowing of the renal artery. This is at present the most accurate diagnostic method.

It must be evident that it is difficult to prove that a specific case of arterial hypertension is caused by narrowing of the renal artery except on aortography and, although aortography is much less dangerous than the average doctor thinks it is, a certain risk is still attached to the procedure so that aortography cannot at this stage be accepted as a legitimate routine investigation in all cases of hypertension.

It is difficult to decide in which cases to do an aortography; the following findings can, however, serve as a guide because they point to narrowing of the renal artery as a causative factor (after the exclusion of other possible causes):

i. The onset of hypertension before the age of thirty years (congenital malformations).
ii. The onset, for the first time, of hypertension after the age of fifty-five years (atherosclerosis).
iii. A sudden exacerbation of existing hypertension.
iv. In cases in which a systolic murmur can be heard in the abdomen.
v. In cases in which a difference in size of the two kidneys cannot be explained on other grounds.

Surgical treatment is often possible provided irreversible changes have not set in as a result of the long duration of the condition. In this connection it is interesting to observe that in unilateral narrowing of the artery, the kidney with a normal blood supply is usually subject to more damage from the hypertension than the kidney with the narrowed artery, because the latter kidney is protected against the raised pressure by the constriction. This is one of the special reasons why an attempt is made to repair the arterial lumen rather than to remove the kidney.

Surgical operations on the renal artery include excision of the stenosis and stitching, lienorenal anastomosis, aortorenal transplantation and endarterectomy. Nephrectomy is indicated in cases in which repair of the vessels is not possible and in which the condition of the other artery is satisfactory. The results are dramatic in suitable cases and it is then permissible to refer to the condition as having been 'cured'. Marked improvement is often achieved in less suitable cases.

Although this condition is seen only rarely at present, it is possible that finer chemical and X-ray investigations might in due course lead to the diagnosis of more cases of this nature. In fact, the possibility of this mechanism operating in branches of the renal artery, and even in the small intrarenal vessels thus causing 'essential' hypertension, is being discussed at present. The proof of these theories is still lacking, but it is of great importance to the patient in whose case a raised blood pressure is caused by narrowing of the renal artery, to have the condition diagnosed and treated surgically.

1. DeCamp, P. T., Birchall, R. and Batson, H. (1958): Scientific Exhibition held at a meeting of the American Medical Association in San Francisco.
2. Goldblatt, H., Lynch, J., Hanzal, R. F. and Summerville, W. W. (1934): J. Exper. Med., 59, 374.

TETANUS NEONATORUM*

R. WRIGHT, M.B., CH.B., *Medical Registrar, Department of Medicine, University of Natal, and King Edward VIII Hospital, Durban*

Despite the high incidence of tetanus neonatorum in South Africa a large series of cases has not been recorded. Friedlander,[1] and Klenerman and Scragg,[2] drew attention to the frequency of the condition in Durban, and there have also been reports from the other large centres.[3-5] During the 3-year period May 1956 to April 1959, 246 cases of tetanus neonatorum were admitted to King Edward VIII Hospital, Durban. Of these, 217, admitted to a special tetanus unit, are the subject of this report.

Race, sex and age. The average age on admission was 7 days. There were 198 African and 19 Indian infants, a ratio of approximately 10 to 1, and males outnumbered females by 129 to 89. The seasonal incidence was not striking, though there was a slight increase during the summer months.

Infection. The organism was rarely isolated, but the umbilicus was always presumed to be the source of infection, though in a few instances it did not appear septic.

Clinical features. The appearance of an established case is unmistakable. The face is pursed up in risus sardonicus and the limbs are partly flexed and stiff, with fists clenched around the thumb and toes plantar-flexed. There is usually some degree of opisthotonus and characteristically stiffness of the abdominal muscles. By placing one hand on the infant's abdomen and the other over the spinal muscles we have found the simultaneous contraction of these antagonistic muscle groups to be a sign pathognomonic of reflex tetanic spasms.

Diagnosis. This picture is so characteristic that we agree with Jelliffe[6] that there is no valid differential diagnosis, and differentiation from conditions causing convulsions in the neonatal period is easy. Some difficulty may occur in the minority of patients (7% in this series) who are not having typical reflex spasms when first seen. Infants suffering from meningitis, birth trauma or sclerema have shown sufficient resemblance to tetanus to be admitted with this diagnosis. Necropsies were performed in all deaths and in none of them was the clinical diagnosis of tetanus disproved.

Treatment. The general principles of treatment remained constant throughout the series. One hour after sedation a single dose of 50,000 international units (i.u.) of antitetanus serum was given intramuscularly. Benethamine penicillin, 300,000 i.u., was injected by the same route and repeated every third day unless signs of pneumonia supervened, when soluble penicillin or a broad-spectrum antibiotic was substituted. Local treatment of the umbilicus was restricted to cleaning with hydrogen peroxide and the application of merthiolate.

Feeding. When spasms had been reasonably well controlled, an intragastric rubber or polyethylene tube was passed by the nasal or oral route and feeds of expressed breast milk given. However, because poor absorption and aspiration of feeds occurred quite commonly, intravenous feeding by intermittent scalp-vein infusion has been recently attempted in a few infants. Most of these were hypothermic and their fluid requirements low, so that overhydration with the development of oedema was a constant danger. Difficulty in providing adequate nutrition during the stage of reflex spasms and rigidity resulted in death from marasmus and bronchopneumonia as late as the 5th or 6th week.

Sedation. Chlorpromazine has been compared with barbiturates and a mixture of barbiturate and chloral hydrate in two random trials which have been reported elsewhere.[7,8] Chlorpromazine (or acetylpromazine, another phenothiazine derivative) was used in combination with a barbiturate in a large number of cases in an attempt to reduce the toxic effects of both drugs. Table I shows the various combinations and the dosage of the drugs used. When once spasms had been controlled, particularly in the phenothiazine-barbiturate groups, the dosage required was usually at the minimum of the daily range.

TABLE I. DRUG COMBINATIONS AND DOSAGES USED

Drug	*Size of Dose (mg. intramuscularly)*	*Daily Range (mg.)*
Chlorpromazine ..	25	100—200
Phenobarb. sod. ..	60	60—300
Phenobarb. sod.* + chloral hydrate ..	120 orally	240—720
Phenobarb. sod. ..	30—60	30—180
+ chlorpromazine	12½—25	12½—100
Phenobarb. sod. ..	30—60	30—180
+ acetylpromazine	5—10	5— 40

*300 mg. (intramuscularly) maximum for 1st 24 hours.

Tracheotomy. Tracheotomy was performed on 17 patients, 13 of whom were compared in a random clinical trial with a conservative method, a mixture of chlorpromazine and barbiturate for sedation being used in both groups.

DISCUSSION

Incidence. It is difficult to compare accurately the incidence of tetanus neonatorum in Durban and district with that elsewhere. Table II has been constructed from reports in the literature to emphasize the alarmingly high incidence at Durban. Table III shows the figures for the past year for neonatal deaths in King Edward VIII Hospital, Durban, indicating that tetanus is one of the major killers. Because of the rapid period of onset and early death if spasms are uncontrolled, it is likely that many more infants die before reaching hospital.

TABLE II. INCIDENCE OF TETANUS NEONATORUM

Area	*Period under Review*	*No. of Deaths or Cases*
Great Britain[9] ..	1938—47	36 deaths
United States[10] ..	1951—55	370 deaths
Singapore[11]	1946—50	254 cases
Ibadan[12]	1953—56	141 cases
Durban	1956—59	246 cases

Aetiological factors. Though no specific custom or method of delivery can be incriminated, about a third of the mothers gave a history of the application to the umbilicus of a 'black powder' obtained from a witch-doctor, and in a few cases *Clostridium tetani* has been cultured from this substance. Confinement on a mat in a hut or shack with an earthen floor, and the use of an unsterilized razor blade, pair of scissors or sharp reed for cutting the cord, has not surprisingly resulted in umbilical sepsis. However, quite frequently the

* Paper presented at the 42nd South African Medical Congress (M.A.S.A.), East London, C.P., September-October 1959.

TABLE III. NEONATAL DEATHS AT KING EDWARD VIII HOSPITAL FOR THE YEAR ENDED 30 JUNE 1959

		%
Prematurity	167	26·2
Tetanus	90	14·2
Asphyxia neonatorum and atelectasis	87	13·2
Bronchopneumonia	79	12·4
Gastro-enteritis	72	11·3
Miscellaneous	61	9·6
Haemorrhagic disorders	49	7·7
Cerebral haemorrhage	18	2·8
Meningitis and septicaemia	12	1·9
Total	635	

delivery took place in a location or domestic servant's quarters, and some of the mothers, occasionally with teaching or nursing experience, had obviously made some attempt at an hygienic confinement, or had even been delivered in hospital.

Prognostic criteria and criteria of severity. Spivey *et al.*[13] have used a 7-day incubation period as the critical level for prognosis. In the present series only 7% of deaths had an incubation period greater than 7 days and only 8% a period of onset of more than 24 hours. While a short incubation period and a short period of onset usually indicate a poor prognosis, as many as 24% of the recoveries had an incubation period of less than 6 days, and 30% a period of onset of under 6 hours. The most useful prognostic sign is the severity of reflex spasms on admission; only 1 of 64 cases in which they were *spontaneous* recovered, whereas 12 of the 15 infants who were not having typical spasms on admission survived. In 8 of these, reflex spasms as described above were never observed, but stiffness and facies were so typical that they are classed in a small group corresponding to mild tetanus in non-neonatal patients. The existence of this type of case, as well as another small group in whom spasms were only moderately severe, though together comprising less than 10% of the series, nevertheless emphasize the need to randomize when comparing different treatment groups.

Complications and mechanism of death. We have found considerable difficulty in deciding on the exact cause of death in the majority of cases. In general terms there would appear to be 4 groups, viz:

1. Uncontrolled spasms, usually in those dying within the first 48 hours, anoxia and exhaustion being largely responsible.
2. Respiratory failure, occurring between the 3rd day and the end of the 2nd week; the action of the toxin on the medullary centres, over-sedation and pulmonary infection and atelectasis being factors in its causation.
3. Marasmus with terminal bronchopneumonia between the 3rd and 8th weeks.
4. A miscellaneous group including tracheo-oesophageal fistula from prolonged tube feeding, neonatal peritonitis and aspiration of feeds.

Necropsies on those dying early usually show acute congestion of the lungs and liver with intra-alveolar haemorrhages and cerebral oedema. Histological evidence of bronchopneumonia was present in 37% of deaths, its incidence being directly proportional to the survival time. Two of the infants who recovered showed radiological evidence of compression of the mid-thoracic vertebrae.

Comparison of sedatives used. Table IV shows the mortality, and the average survival time, in those dying within 14 days in the different treatment groups. Experimentally the phenothiazine derivatives have been shown by Laurence and Webster[14] to have a potent antitetanic action in animals, and this has been confirmed clinically in non-neonatal tetanus.[7,8]

TABLE IV. COMPARISON OF DIFFERENT TREATMENT GROUPS

No. of Cases	*Treatment*	*Percentage Mortality*	*Survival Time* (days)*
17	Barbiturate	72	2·2
34	Chlorpromazine	94	2·0
20	Barbiturate + chloral hydrate	90	3·8
77	Chlorpromazine + barbiturate	76	4·0
34	Acetylpromazine + barbiturate	74	5·2
17	Tracheotomy	100	4·1
18	Miscellaneous	92	
217	(Recoveries 38)	82·5	

* Average survival time in deaths under 14 days.

In two clinical trials in the present series phenothiazine derivatives used alone have not been effective in controlling reflex spasms in 75% of cases, even if given in large doses, and the mortality has been high and the survival time short. In this respect we have not been able to confirm the findings of other workers.[15,16] When they have been combined with barbiturates, spasms have been controlled in all but 30% of cases, with some reduction in mortality, but in general death from uncontrolled spasms within the first 48 hours has been replaced by death from respiratory failure a few days later. In Table V our average mortality throughout the series is compared with that in other large series published recently. With one surprising exception,[20] in which full details of cases are not given, the similarity of the results with different conservative methods of treatment suggests that there is little to choose between the sedatives used.

TABLE V. COMPARISON OF MORTALITY WITH THAT IN OTHER LARGE SERIES

Series	*No. of Cases*	*Sedative used*	*Percentage Mortality*
Present series	217	acetylpromazine chlorpromazine barbiturate etc.	82·5
Jelliff *et al.*[17]	26	barbiturate	96
Spivey[13]	25	paraldehyde chloral hydrate	77
Loh Siew Gek[11]	174	paraldehyde chloral hydrate	92
Sarrouy *et al.*[18]	20	chlorpromazine barbiturate relaxant	80
Pinheiro[19]	256	barbiturate chloral hydrate myanesin	84
Tompkins[12]	141	paraldehyde chloral hydrate barbiturate	89·6
Earle *et al.*[20]	32	barbiturate chlorpromazine	25

In an attempt to reduce the mortality from respiratory failure, a random trial was conducted to assess the value of tracheotomy in preventing pneumonia and anoxia. The sedative used was a barbiturate-chlorpromazine combination and tracheotomy was performed under local anaesthesia, a metal tube being inserted through a window cut in the trachea. Oxygen was administered when required by means of a funnel or catheter but without artificial respiration or humidification of the inspired air. Only very severe cases were selected for

trial. Though the mortality was 100% in both groups, the survival time in the conservatively treated group was longer than in those on whom tracheotomy was performed, two cases in the former dying of late complications at 42 and 37 days respectively. It is apparent that some form of assisted respiration is necessary in the treatment or prevention of respiratory failure, and a trial is now in progress of total curarization, tracheotomy, and intermittent positive-pressure respiration. The constant medical and nursing attention required may alone be an important factor in reducing mortality, making it essential to randomize when assessing the value of this form of treatment. While our own and other preliminary findings[5] indicate a striking increase in survival time, care should be taken in confining such a radical procedure to severe cases; even if it is shown to be effective its use on a large scale will be limited by the expense and the need for specially trained personnel.

Preventive treatment. The only practical method of reducing the high death rate from this disease lies in its prevention. While this can only come with radical changes in the educational and socio-economic status of our African and Indian populations, a few measures may be advocated under present circumstances. A campaign by the local authorities should be directed against the current tribal customs of applying foreign material to the cord, and advice given on simple methods of hygiene at the time of delivery. Active immunization of mothers during pregnancy in an attempt to produce protective antibody levels in their newborn infants has been suggested,[21] and transplacental transmission has been demonstrated experimentally.[22] At least 2 inoculations are given at an interval of 6 weeks during pregnancy. Mothers delivered in hospital should be carefully instructed in the care of the cord; the development of tetanus in hospital-born infants who are discharged prematurely has been well documented,[10,13] and has occurred in this series. Lastly, in view of the high incidence and appalling mortality from the disease, it may well be that the administration of prophylactic tetanus antitoxin, which has become a routine in the treatment of surgical wounds, is as strongly indicated in newborn infants at risk in areas where tetanus neonatorum is endemic.

SUMMARY

Over 200 cases of neonatal tetanus have been admitted to a special unit of the Department of Medicine, King Edward VIII Hospital, Durban, during a 3-year period. The alarmingly high incidence of the disease in the area is emphasized, and possible aetiological factors mentioned. The diagnosis, clinical features, prognostic criteria and mechanism of death in these cases are discussed. Various forms of therapy are described, some of which were investigated in randomized clinical trials, and the value of phenothiazine derivatives and other drugs in the suppression of tetanic spasms compared. The literature is briefly reviewed and the problems of therapy discussed, with special reference to the control of reflex spasms and the treatment and prevention of respiratory failure.

In view of the high mortality, public-health measures are urged in an attempt to prevent the disease.

I wish to thank Prof. E. B. Adams for his encouragement and guidance; Dr. N. M. Mann for advice on feeding problems and assistance with fluid therapy; Mr. D. R. Gowans, Mr. G. Immerman and Mr. D. C. Carter for performing the tracheotomies; Dr. S. Disler, Medical Superintendent, for facilities; and Matron Uijs and the members of the nursing staff for their cooperation.

The Tetanus Research Unit has been supported by grants from the Wellcome Trust.

REFERENCES

1. Friedlander, F. C. (1951): J. Pediat., **39**, 448.
2. Klenerman, P. and Scragg, J. (1955): S. Afr. Med. J., **29**, 853.
3. Slome, R. (1954): *Ibid.*, **28**, 473.
4. Falcke, H. C. (1957): Med. Proc., **3**, 171.
5. Smythe, P. M. and Bull, A. (1959): Brit.Med. J., **2**, 107.
6. Trowell, H. C. and Jelliffe, D. B. (1958): *Diseases of Childhood in Subtropics and Tropics*, 1st ed., p. 104. London: Arnold.
7. Laurence, D. R., Berman, E., Scragg, J. N. and Adams, E. B. (1958): Lancet, **1**, 987.
8. Adams, E. B., Wright, R., Berman, E. and Laurence, D. R. (1959): *Ibid.*, **1**, 755.
9. Conybeare, E. T. and Logan, N. P. D. (1951): Brit. Med. J., **1**, 504.
10. Axnick, N. W. and Alexander, E. R. (1957): Amer. J. Publ. Hlth, **47**, 1493.
11. Loh Siew Gek (1951): Med. J. Malaya, **5**, 181.
12. Tompkins, A. B. (1958): Brit. Med. J., **1**, 1382.
13. Spivey, O. S., Grulee, C. G. and Hickman, B. T. (1953): J. Pediat., **42**, 345.
14. Laurence, D. R. and Webster, R. A. (1958): Brit. J. Pharmacol., **13**, 334.
15. Gelfand, M. (1955): Cent. Afr. J. Med., **1**, 216.
16. *Idem* (1957): *Ibid.*, **1**, 90.
17. Jelliffe, D. B. (1950): Arch. Dis. Childh., **25**, 190.
18. Sarrouy, C., Gillot, F., Clausse, J., De Peritti, E. and Gatto, L. (1956): Algérie Méd., **60**, 277.
19. Pinheiro, D. (1957): J. Pediat., **51**, 171.
20. Earle, A. M. and Mellon, W. L. (1958): Amer. J. Trop. Med. Hyg., **7**, 315.
21. Rogers, L. (1944): Indian Med. Gaz., **59**, 297.
22. Ten Broeck, C. and Bauer, J. H. (1922-23): Proc. Soc. Exp. Biol. (N.Y.), **20**, 399.

PHENYLKETONURIA: REPORT AND DISCUSSION OF THREE CASES

WALTER KLUGE, M.B., CH.B., *Alexandra Institution, Maitland, Cape*

During more recent years biochemical methods of investigation have enabled us to gain a deeper insight into the nature of certain types of defective mental development. This work signifies a comparatively new approach towards a better understanding and even a possible solution of some of the many hidden problems of mental retardation. In this manner a few cases of defective mental development have been crystallized out of the large group of primary amentia and have been found to be associated with or even caused by errors of metabolism of a specific nutrient or substrate. Work on these novel lines continues and will no doubt throw more light on conditions of mental defect.

In accordance with the major metabolic substrate involved, these disorders have been classified under the following headings:[1,2]

1. Errors of protein metabolism represented by phenylketonuria, H-disease, and hepatolenticular degeneration (Wilson's disease).

2. Errors of carbohydrate metabolism represented by galactosaemia, idiopathic hypoglycaemia, and gargoylism.

3. Errors of fat metabolism represented by the group of familial amaurotic idiocies, Niemann-Pick's disease, and Gaucher's disease.

4. Disturbances of hormone synthesis represented by familial sporadic cretinism with goitre.

5. Cases of probable defective metabolism such as idiopathic hypercalcaemia of the infant, Lowe's disease, pitressin-resistive diabetes insipidus, and congenital familial non-haemolytic jaundice.

On none of these types of mental deficiency accompanied or caused by metabolic anomalies has more attention been focussed than on phenylketonuria, probably because of its easy mode of diagnosis by a simple urine test, because of the hope it holds forth of responding to treatment, and because it is the most frequent and best defined representative of this group of metabolic syndromes.

About 7 years ago, Dr. E. H. Welsh, then a member of the medical staff of the Alexandra Institution for the Feebleminded, and Sister van Zyl, now assistant matron at the institution, conducted a preliminary investigation into the existence of phenylketonuria among the inmates, and found only one case. Recently I undertook a systematic search for this condition among 888 patients of the same institution (including 707 White cases of mental deficiency and 181 non-White mentally disordered patients with psychoses such as schizophrenia, manic depressive and epileptic psychoses and dementia paralytica); 3 cases of phenylketonuria were found among the 707 mentally defective patients, and none among the psychotic patients. A possible 4th case was the clinically suspected familial case who had been a patient here for many years until her death 8 years ago.

As far as I know only one case of phenylketonuria has hitherto been recorded in the South African medical literature (Braude[3]).

CASE RECORDS

First Case

The strange fact that the urine of this patient, the very first of all the 888 urines examined for this investigation, should have been positive for phenylketonuria, is not entirely due to coincidence, for he is one of a group of 4 patients whom, for certain physical features present in them, I had selected as likely cases of this condition.

He was admitted to this institution as a certified mentally defective patient on 23 June 1946 at the age of 3 years. His weight was then 36 lb. (normal average 31½ lb.), his height 41 inches (normal average 35 inches), and his state of nourishment good; he was well developed and all his tendon reflexes were within normal limits, but his gait was described as slightly uncertain. His WR was negative.

Unfortunately a proper history of his case has been unprocurable. As far as can be ascertained there is no mental abnormality among members of his family. The first note in his case-sheet, dated 24 June 1946, is instructive: 'His appearance is not unpleasant, he is quite well formed, has fair hair and blue eyes and lacks any of those signs characteristic of special forms of amentia. He is clearly mentally backward for a child of his age, is emotionally unstable and cries and screams on very slight provocation; his vocabulary is limited to 'Papa' and 'Mama'. He has outbursts of uncontrollable, violent temper, when he mutilates himself and hurts others within reach. He is almost completely helpless and nearly everything must be done for him.' On admission, and for 13 years thereafter his condition had been diagnosed as primary amentia, simple type, idiocy (according to the classification of Tredgold sr.). The examiner was obviously puzzled by the patient's pleasant appearance, blue eyes and fair hair. Until fairly recently no attention had been paid to phenylketonuria at this institution, and the possibility that this patient was a phenylketonuric had therefore not been considered or even vaguely contemplated. During the 13 years of his hospitalization his mental condition has remained stationary, and his conduct as unruly as ever. His intestines were repeatedly infested with round worms, and he often had most obstinate attacks of microscopically confirmed amoebic dysentery.

At present he is 16 years old, his weight 81 lb. (normal average 123¾ lb.), and his height 60 inches (normal average 65¾ inches). Even the senior nursing staff are impressed by his white skin, blue eyes and fair hair. Brown marks, the size of a sixpence to a shilling, which he has in his skin on the back of his hands and on his legs, are attributed to blisters; in his case-sheet notes he is described as 'rather subject to sunburn'. He has healthy and normally spaced and aligned teeth. The tendon reflexes of his upper extremities are brisk, those of his lower extremities very brisk. He has moderate talipes varus. As the result of the deformity of his feet, and probably also of the increased tendon reflexes of his lower extremities, his gait is slightly impaired and suggestive of spasticity. His posture has a tendency to flexion. He is so constantly restless, that he is on the move nearly all the time, either running about wildly and aimlessly or performing the same movements over and over again in rhythmic fashion, such as rocking his trunk backwards and forwards when in a sitting position, bouncing a big rubber ball and each time with remarkable skill jumping high to catch it without fail, or suddenly spinning his body round its long axis, etc. He is a typical example of phenylketonuria.

Second and Third Cases

As these two cases, both females, are also fairly typical instances, and their full description would in some respects be just a repetition of the preceding, only the essential points of their condition will be mentioned. There is nothing in their case-histories to throw light on a possible cause of their disorders. They are both certified mental defectives, the one with an intelligence level of feeblemindedness (IQ 67), the other an idiot with IQ 22. They both have been in this institution for many years.

The case of feeblemindedness, now 47 years of age, is a quiet, reserved, very good and pleasant, hard-working person, who spends much of her time attending to the helpless low-grade patients. She has an attractive appearance, with fair hair, blue eyes and a delicate skin. Her late sister, an imbecile, probably also a phenylketonuric, was a patient here.

The other patient of our series is a very restless, resistive, apprehensive, highly tensed, helpless idiot. She too has fair hair, very blue eyes and a delicate skin. As a baby she had bad eczema of the face, and her father is hypersensitive to sun-rays. She is a very healthy, strapping, pretty girl. Her mother's urine reacts normally to the ferric-chloride test.

All 3 of our cases had until quite recently been classified as instances of simple primary amentia only, without further specification, until by the application of the ferric-chloride test to their urines, they were found to be typical phenylketonurics.

DISCUSSION

The diagnosis of simple primary amentia which constitutes about 70% of all cases of mental deficiency and which sometimes is arrived at only by a process of exclusion did not always satisfy. It was felt that in some of these cases there must be another factor involved, apart from the genetically determined abnormality of the germ plasm. Continued investigations shed some light on this problem, when in 1934 Følling, a Norwegian biochemist, found that the urines of certain low-grade cases of mental deficiency developed a green colour on the addition of ferric-chloride solution. This reaction is caused by the presence of phenylpyruvic acid, which is the breakdown product of the amino-acid phenylalanine, and normally is not present in the urine. Følling named those cases in which the urine behaved in this manner 'imbecilitas phenylpyruvica'.

His observations were soon confirmed by others, and Penrose named the condition phenylketonuria; it is also known as phenylpyruvic amentia or oligophrenia. Phenylketonuria is a variety of simple primary amentia associated with or, as some believe, actually caused by an error of protein metabolism. The disorder is produced by a recessively inherited inability, possibly of the liver, to supply a sufficiency

of the enzyme that is necessary for the oxidation of the amino-acid phenylalanine, which in the normal person is almost entirely converted into tyrosine. In the phenylketonuric the blocking of this process results in the accumulation in the blood, tissues and cerebrospinal fluid, of phenylalanine and its products of incomplete inoxidation such as phenylpyruvic acid, phenyllactic acid, phenylacetic acid and others, which are considered to have a toxic effect on the functions of organs such as the brain. Others again believe that phenylketonuria is caused by the inability of the tissues to utilize certain metabolites.

Phenylketonuria usually presents no special clinical manifestations by which it can be distinguished from the ordinary simple variety of primary amentia. Occasionally, however, its diagnosis can be suspected by the presence of an assemblage of fairly characteristic physical features, sometimes, it is claimed, aided by an aromatic odour given off by its victims and their urine; but the presence of phenylketonuria can be conclusively determined only by the application of chemical tests to the urine and blood.

The test generally used, because it is so simple, easily performed and at the same time highly sensitive, is to add about 10 drops of a 5% solution of ferric chloride to about 5 c.c. of urine acidified with a few drops of dilute sulphuric acid; if the urine develops a light apple-green colour which rapidly turns deep-green, then the diagnosis of phenylketonuria is confirmed. In cases where a sample of urine is unobtainable, as with babies, we make use of the diaper test,[4] in which a drop of a 5% ferric-chloride solution is placed on the urine-soaked diaper; the test is positive for phenylketonuria if a grey-green or blue-green spot appears, and negative if the spot is yellow. In this investigation the diaper test was modified by first applying a drop of dilute sulphuric acid and then on top of it the drop of ferric-chloride solution. The results of these tests were all so convincingly negative that, in spite of recently raised doubts about the reliability of the test, there did not exist the slightest reason to question the findings in the 41 cases on which it was done in the investigation.

Apparently, by means of the phenylalanine tolerance test and the phenylalanine-tyrosine ratio, one is able to detect heterozygous carriers.[5]

As phenylketonuria and its abnormal constituents of the urine are present at birth,[6] and as it is one of the few instances of mental deficiency where medical treatment actually holds forth some hope—but only if it is tackled thoroughly and early enough—and as its diagnosis can be confirmed so easily by means of the ferric-chloride urine test, it is most important to perform this test on the urine of every newborn child as soon as possible after its birth. It should be repeated several times at intervals of a few weeks to check the findings of the previous urine tests and to control the effect of dietary treatment if this has been instituted; and also because there have been instances where phenylpyruvic acid has not been persistently present in the urine, although in most cases it is excreted constantly.[17] Recent investigations conducted on affected siblings of known cases of phenylketonuria appear to indicate that the blood level of phenylalanine begins to rise between the 2nd and 6th week of life, and therefore it is recommended that the urine should be tested for phenylketones at the age of 3 weeks and again at 6 weeks, and treatment started at about the same time, if positive.[8,9]

If the result of the urine test is in any way doubtful, the case should be referred to a biochemical laboratory for a confirmatory test with dinitrophenylhydrazine, and preferably also a quantitative estimate made of the phenylpyruvic acid in the urine and of the phenylalanine in the serum.

Levy and Perry[9] found the incidence of phenylketonuria to be 0·693% among the intellectually retarded they had examined.[10] Jervis found a total of 161 cases among 20,300 defectives in various institutions of the USA.[11] Phenylketonuria occurs about once in 25,000 births.[2] As stated above, at the Alexandra Institution 3 cases of this condition were traced among the 707 mentally defective inmates (0·43%) and none among the 181 non-White mentally disordered patients.

Evidence of hereditary origin is present only in one of the cases of phenylketonuria; her late sister, a certified mental defective of imbecile level of intelligence was an inmate of the same institution and, although the pathognomonic ferric-chloride urine test had not been performed, yet sufficient clinical evidence was present (such as blond hair, blue eyes, fair delicate skin and athetoid movements) to make her condition suggestive of phenylketonuria.

The grade of intelligence of phenylketonuria is usually not higher than that of imbecility (in about 70% of the cases) or idiocy (in about 30% of the cases),[10] although more recently a few instances, probably not exceeding 0·1%,[12] with an intelligence approaching the normal range, have been reported.[12-15] Among the cases traced here, one is feeble-minded (IQ 64) and the other two are idiots (IQ 22 and 4 respectively).

In phenylketonuria there seems to exist a connection or correlation between the degree of metabolic error, i.e. the total daily output of phenylpyruvic acid, and the severity of mental defect.[12]

A fairly well defined constitutional body make-up was found in a large number of patients afflicted with phenylketonuria, in some instances sufficiently pronounced to suggest its diagnosis. About 77% have blue eyes, fair hair and hyperactive tendon reflexes.[9] Our 3 cases are all characterized by this dilution of the colour of their eyes, hair and skin and by neurological manifestations like brisk tendon reflexes. One case displayed spasmodic contractions of the head; another had athetoid movements. All these neurological manifestations are attributable to lesions of the extrapyramidal system.[10] They all have pleasant, attractive facial features. With the exception of deformed feet in one instance, all 3 cases of our series are physically well developed and very healthy.

In none of them has there been any evidence of mental deterioration during many years. Investigations seem to indicate that the intelligence of most phenylketonurics deteriorates rapidly during the first months of life, so that at about 12 months a state of imbecility or idiocy is usually reached, and thereafter mental deterioration is only very slow.[9]

As mentioned above, phenylketonuria is one of the very few conditions of mental deficiency the others are those associated with cretinism, galactosaemia and ideopathic hypoglycaemia, with the very remote possibility also of mongolism, —which, as recent investigations have shown, actually hold forth hope of being amenable to treatment if carried out efficiently and early enough to forestall irreversible damage to the brain. The treatment is to put the patient on a

phenylalanine- and tryptophan-restricted diet. In a few cases of phenylketonuria which were recently treated in this manner very pleasing results were obtained. A list of references to this dietary treatment is given at the end of this article.

That in addition to the knowledge, although still very fragmentary, which we have of the prevention of certain types of mental deficiency, we can now actually apply medical treatment, with hopeful results, to a few varieties of defective mental development, is a remarkable achievement, particularly in view of the firmly fixed attitude of defeatism and negativism hitherto generally adopted. This new development, which is still a faint glimmer on the distant horizon, is bound to stimulate continued and still more intensified efforts in this direction.

SUMMARY

Three cases of phenylketonuria are described and discussed.

OPSOMMING

Drie gevalle van fenielketonuria word beskryf en bespreek.

I wish to thank Dr. B. P. Pienaar, Commissioner for Mental Hygiene, and Dr. M. Cohen, Physician Superintendent of the Alexandra Institution, for permission to submit this article for publication. I am particularly indebted to Dr. H.W. Smith, Psychologist to the Alexandra Institution, for most valuable help and advice.

REFERENCES

1. Wright, W. *et al.* (1958): Amer. J. Dis. Child., **95**, 541.
2. Jochims, J. (1958): Med. Klin., **42**, 1835.
3. Braude, H. (1956): S. Afr. Med. J., **30**, 83.
4. Abbott Laboratories, *What's New*, no. 210, p. 7.
5. (*a*) Hsia, D. Y. Y. *et al.* (1956): Lancet, **2**, 1337.
5. (*b*) Hsia, D. Y. Y. *et al.* (1957): J. Ment. Defic. Res., **1**, 53.
5. (*c*) Hsia, D. Y. Y. (1958): *Ibid.*, **2**, 8.
6. Hilliard, L. T. and Kirman, B. H. (1957): *Mental Deficiency*, p. 335. London: Churchill.
7. Follings, A. *et al.* (1945): Quoted by Hilliard *et al.*, *op cit.*[6]
8. Armstrong, M. D. and Buckley, E. L. (1956): Proc. Soc. Exp. Biol. (N.Y.), **93**, 418. Quoted by Brimblecombe *et al.*, *loc. cit.*[9]
9. Brimblecombe, F. S. W. *et al.* (1959): Lancet, **1**, 611.
10. Levy, S. *et al.* (1949): Amer. J. Ment. Defic., **54**, 73.
11. Tredgold, R. F. and Soddy, K. (1956): *Text-book of Mental Deficiency*, 9th ed., p. 291. London: Baillière, Tindall and Cox.
12. Cowie, V. *et al.* (1958): J. Ment. Defic. Res., **2**, 55.
13. Low, N. L. *et al.* (1956): Lancet, **2**, 917.
14. Coates, S. *et al.* (1957): Arch. Dis. Childh., **32**, 313.
15. Hsia, D. Y. Y. *et al.* (1957): Amer. J. Dis. Child., **94**, 33.

REFERENCES ON DIETARY TREATMENT

Bickel, H. *et al.* (1954): Acta paediat., **43**, 65.
Woolf, L. I. *et al.* (1955): Brit. Med. J., **1**, 57.
Armstrong, M. D. and Tyler, F. H. (1955): J. Clin. Invest., **34**, 565.
Braude, H. (1956): S. Afr. Med. J., **30**, 83.
Horner, F. A. *et al.* (1956): J. Amer. Med. Assoc., **161**, 1628.
Hsia, D. Y. Y. *et al.* (1958): Pediatrics, **21**, 178.
Brimblecombe, F. S. W. *et al.* (1959): Lancet, **1**, 611.

* Baron, N. D. *et al.* (1956): Lancet, **2**, 421.
* Lang, K. *et al.* (1938): Z. Kinderheilk., **80**, 311.
* Opitz, H. and de Rudder, B. (1957): *Paediatrie*, in chapter by Hövels *et al.* Berlin: Springer Verlag.
* Schultze, S. (1958): Mschr. Kinderheilk., **106**, 203.
* Stanbury, J. B. *et al.* (1957): Amer. J. Med., **22**, 712.

* References kindly supplied by Prof. Dr. Med. J. Jochims, Chefarzt der Städt Kinderklinik, Lübeck, Germany.

ENGELMANN'S DISEASE: CASE REPORT AND BRIEF REVIEW

I. M. PATZ, M.B., B.CH. (RAND), *Middelburg, Transvaal*, and H. J. J. VAN HEERDEN, M.B., B.CH. (RAND), D.M.R.D. (LOND.),* *Germiston, Transvaal*

Engelmann's disease is a rare clinical entity of which only 23 authenticated cases have been published. It is characterized by sclerosis of the long bones involving chiefly the middle thirds of the shafts of these bones. We present here what we believe to be the 24th case described, illustrating the typical radiological features of the disease along with some special findings.

Engelmann's disease, or progressive diaphyseal dysplasia, was first described by Cumarati[1] in 1922, 7 years before Engelmann's account[2] appeared in print. Griffiths,[3] in an excellent review of the condition (1956), recognized 21 definite cases. Of these 16 were reported in detail, and Griffiths added the detailed account of a further case; the other 5 cases, although more briefly described were sufficiently documented to make the diagnosis certain. A further case has since been described by Stewart and Cole.[4]

CASE REPORT

In March 1955 a 27-year-old unmarried woman was referred for a dysenteric condition of 18 months' duration. She had frequent loose stools daily, containing much blood and mucus. On full investigation, which included repeated stool examinations, roentgenography of the whole gastro-intestinal tract and repeated sigmoidoscopic examinations, and taking into account the patient's psychological background, it was decided that this was a case of chronic non-specific ulcerative colitis, the precipitating factor being an unhappy life situation.

The patient stated that at the age of 7 years she was confined to bed for several months (without medical attention) for an illness of which she could not recollect the details. On recovery from this illness she had difficulty in walking and over the years she developed a peculiar gait. Before this she had walked normally but had always been a thin and ailing child and often suffered from pain in the limbs. Her left leg became shorter than the right, causing a pronounced limp, and the muscles of the lower limbs were weak. As a result of her disablement she left school at the age of 14 and has never done remunerative work, but has been drawing a government disablement pension since the age of 18 years. About 6 months before the onset of the diarrhoea the pension authorities sent her to a large provincial hospital to ascertain the nature of her disability. An X-ray survey of her skeleton revealed 'a peculiar bone disease' (the patient's own words). The patient further stated that she had never menstruated and had no breast development, and that there was complete absence of libido or interest in the opposite sex. As a result of her disabilities she had the feeling that she was 'different to other women' and was constantly being stared at. She has always been thin and although she has a good appetite she has never been able to gain in weight even before the onset of the diarrhoea. In June 1955 her teeth were extracted in the hope that this would improve the diarrhoea. She is one of 10 children and all the others are in good health. There is no history of a similar ailment in the family.

On examination she was found to be tall (5 feet 9 inches) and very thin (Fig. 1). She walked with a waddling gait and a distinct limp to the left. She appeared to have long limbs, the thighs and forearms being especially long. The forehead was high and wide and the scalp hair fine and blond. There was a slight staring appearance of the eyes but no exophthalmos. The fields of vision and the ocular fundi were normal. The facial skin was smooth and soft and there was slight brownish pigmentation of the forehead, cheeks and bridge of the nose. The conjunctival and oral mucous membranes were pale. The pubic and axillary hair was scanty. The heart and lungs were normal on clinical examination. The blood pressure was 115/70 mm. Hg. There was marked tenderness over the whole of the colon, especially the descending colon, which was easily palpable.

* Formerly Department of Radiology, Pretoria General Hospital, University of Pretoria.

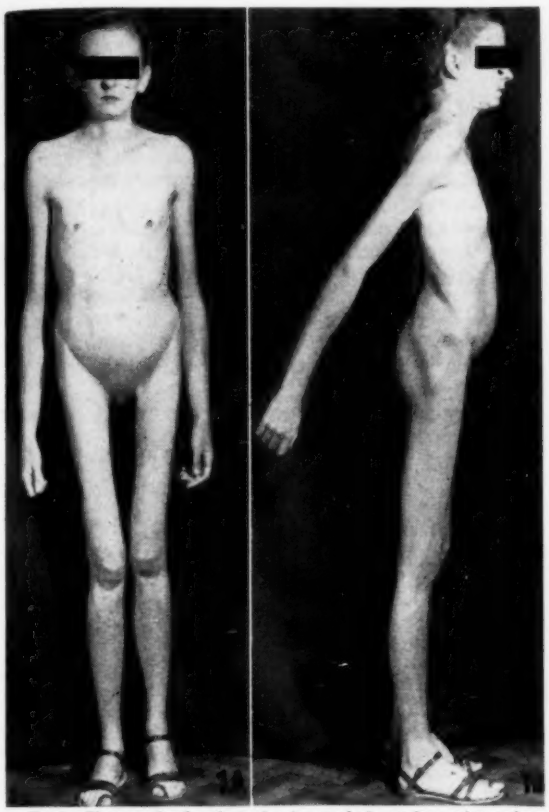


Fig. 1. Photograph of patient, showing marked muscle wasting and absence of breast development.

The breasts were not developed at all. Except for the scantiness of the pubic hair the vulva was normal. The vagina was short and a very small uterus was palpable on vaginal examination. A mass about 1 inch in diameter was felt in the left adnexal area, probably an ovary.

The bones of the arms, forearms, thighs and legs felt thickened. The limb musculature was poorly developed. There was marked limitations of abduction at both hip joints but no limitation in flexion or extension. There was no limitation of movement at the other joints. The tendon reflexes were normal and no disturbance of sensation or other neurological abnormality was found.

Measurements. Length 69 inches, weight 90 lb., pubic height 36 inches, span 73 inches, chest circumference at nipple line 28½ inches, circumference at hips 28 inches, skull diameter 23 inches.

Laboratory findings. Urine analysis was normal. A blood count on 16 March 1955 revealed a severe hypochromic anaemia: Hb. 7·4 g. %, r.b.c. 2,900,000 per c.mm., w.b.c. 5,200 per c.mm. (44% polymorphonuclear leucocytes and 55% lymphocytes). Platelets present in normal numbers. Intramuscular iron therapy (Imferon) was followed by a dramatic improvement in the anaemia; barely 3 weeks after treatment was begun the Hb. rose to 12 g.% and the r.b.c. to 4,000,000 per c.mm., indicating that there was no bone-marrow disturbance causing the anaemia. The modified Ide test yielded a negative result. Chemical analysis yielded the following results: Serum calcium 5·8 mEq./litre; serum sodium 139 mEq./litre; serum potasium 4·1 mEq./litre; blood urea 21 mg./100 ml.; serum albumin 2·3 g.%; serum globulin 2·8 g.%, alkaline phosphatase 13·9 K.A. units. The liver function battery of tests showed equivocal results for liver damage. The sugar tolerance test yielded a normal result. Estimation of 17-keto-steroids in 24-hour urine specimen yielded 3·3 mg. and 2·2 mg.

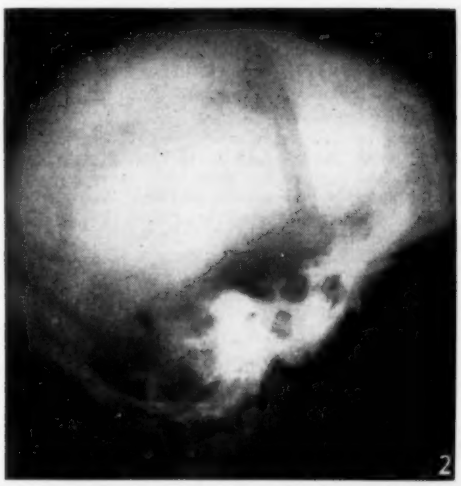


Fig. 2. The skull showing sclerosis of vault and base and involvement of parietal bones.

Fig. 3. The humerus showing sclerosis of the shaft and non-involvement of the metaphyses. The medullary cavity is relatively normal. The other humerus shows the same changes.
Fig. 4. Radius and ulna, demonstrating the sclerosis of the shafts. The other radius and ulna show the same changes.
Fig. 5. The femur showing the sclerosis and thickening of the diaphysis. The other femur shows the same changes.
Fig. 6. The tibia and fibula showing the typical changes. The other tibia and fibula show the same changes.

on separate occasions, and 2·1 mg. of 17-hydroxyethiosteroid (estimated as free cortisone). Estimation of follicle-stimulating hormone yielded more than 6 and less than 12 mouse units in a 24-hour urine specimen. Histological examination of a specimen of skin revealed a female chromosomal pattern.

Radiological Examination

There was gross thickening of the vault of the skull, including the parietal bones and the base of the skull; the squamous temporal bones, basi-occiput and sella were normal (Fig. 2). The facial bones and mandible showed no bony changes.

There was symmetrical expansion of the shafts of the humeri, showing gross cortical thickening. The lower ends of the humeri showed no involvement and the metaphyseal and epiphyseal regions were normal. The medullary cavities were relatively uninvolved (Fig. 3). Similar appearances were seen affecting both radii and ulnae (Fig. 4).

Symmetrical and cylindrical expansion was also seen in both femora (Fig. 5). At the metaphyseal region of the femoral necks a dense sclerotic band was present. The femoral heads, however, were normal (Fig. 7). The upper two-thirds of the fibulae and tibiae were similarly affected, leaving the distal thirds quite normal in appearance (Fig. 7). The spine, pelvis, clavicles, hands and feet showed no bony changes.

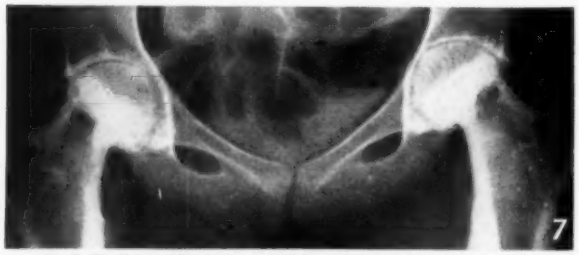


Fig. 7. The femoral heads are normal, but a dense band is seen at junction of head and neck of each femur.

Changes in the femoral necks have only once been reported, by Griffiths.[3] No other case showing the narrow sclerotic band at the junction of the head and neck of the femur could be traced. This case also differs from previously reported cases in that the parietal bones of the skull are also affected.

A barium enema confirmed the presence of ulcerative colitis.

DISCUSSION

Engelmann's disease is characterized by symmetrical enlargement and sclerosis involving the shafts of the major long bones, associated in most cases with skull changes. The changes in the long bones are restricted to the diaphysis, usually the middle two-fourths. According to Griffiths[3] there were skull changes in all but 8 of the 22 cases reviewed by him, with an increased density of the base or vault or both, the changes in the vault involving the frontal bones mainly. Changes were not reported in the parietal bones or the squamous parts of the temporal bones, nor in the facial bones and jaws.[3]

The long bones most often affected are the femora, tibiae, humeri, radii, ulnae, and fibulae. The femora were involved in all cases reviewed by Griffiths[3] and it was these that exhibited the most striking changes. The tibiae were affected in all but 2 of the reported cases. If the tibiae were affected then the fibulae were generally affected as well. The humeri and forearm bones were also involved in most cases.[3] The spine has been reported to be affected in only 1 case,[5] (and then only the atlas was involved), the pelvis in 1 case,[5] the clavicles in 5 cases,[3] and the ribs in 1 case. No other bones have been reported to be involved.

The sex incidence of the disease is about equal, and no case has been described in a negro or in a patient from the Asiatic mainland.[3] The youngest age at which the diagnosis was made was 33 months,[6] and the oldest 55 years.[1] In all adult cases puberty was delayed, the genitalia and secondary sexual characteristics never being well developed. One male adult described as a case of the disease by Stronge and McDowell[7] was sexually normal. Small atrophic testicles have been described in some cases.

Affected children are late walkers and most cases complain of pain in the bones. Most patients are poorly developed and short of stature, the limbs appearing abnormally long in proportion to the height. Postural defects are common and in all cases the gait is described as abnormal and generally of the waddling type. In some cases the abnormality in gait commenced in later childhood. Muscle development is remarkably poor in most cases and the thickened long bones may be palpated in some cases through the thin muscle masses of the extremities. Limitation of hip and knee movements may be present but generally joint movement is free. Investigation of the blood count and blood chemistry have shown no specific or significant deviations. The histology of the involved bone reveals a non-specific picture of osteosclerosis. No evidence of inflammatory changes have been found.

The case described here is presented as one of Engelmann's disease on clinical and radiological grounds. The case presents as a physically underdeveloped female with marked muscle weakness of the extremities, marked muscle wasting, abnormal gait, failure of development of secondary sexual characteristics, and thickened and sclerotic long bones. Radiologically the long bones show the characteristic diaphyseal hyperostosis and the skull the basal and frontal sclerosis. The patient differs from most cases in being tall, only Stronge and McDowell's case[7] also being tall. It is felt that the diarrhoea has nothing to do with the bone condition, which was discovered before to the onset of the diarrhoea. Only in one case, presented for diagnosis by Cockayne[8] in 1920, was looseness of the stools mentioned; Fairbank[9] diagnosed this case as one of Engelmann's disease in later years.

The aetiology of the condition is unknown. Griffiths[3] favours a genetic origin in view of the early onset of the disease, the absence of abnormal cell structure, and the symmetrical progress in the skeleton. It is interesting that Cockayne,[8] even before his case had been given a specific diagnosis, entertained the possibility of an endocrine disorder as being the underlying cause. In view of the late onset of puberty and menstruation in female cases, the presence of small testicles in some of the males, and failure in full development of secondary sexual characteristics in most cases, it is felt that an endocrine disturbance is a possibility as a cause for this condition. It is to be noted that Engelmann's disease is not only a bone disorder but a constitutional disease as characterized by disturbance in general physical and sexual development.

SUMMARY

A case of Engelmann's disease (progressive diaphyseal dysplasia) is presented, with a brief survey of the clinical and radiological features.

We wish to thank the Superintendent of the Middelburg Hospital for permission to publish this case. All the laboratory investiga-

tions were carried out by the South African Institute for Medical Research, Johannesburg. Dr. S. Lopis, Johannesburg, kindly supervised the endocrinological investigations. We wish to record our thanks to Prof. S. F. Oosthuizen and Dr. T. Fichardt, of the Department of Radiology, Pretoria Hospital, for their help and guidance in the preparation of the case for publication.

REFERENCES

1. Cumerati, M. (1922): Chir. Organi Mov., **6**, 662. Quoted by Griffiths, *loc.cit.*[3]
2. Engelmann, G. (1929): Fortschr. Röntgenstr., **39**, 1101.
3. Griffiths, D. L. (1956): J. Bone Jt Surg., **38B**, 312.
4. Stewart, H. B. and Cole, E. R. (1956): J. Pediat., **48**, 482.
5. Sear, H. R. (1948): Brit. J. Radiol., **21**, 236.
6. Gillespie, J. B. and Mussey, R. D. (1951): J. Pediat., **38**, 55.
7. Stronge, R. F. and McDowell, H. B. (1950): J. Bone Jt Surg., **32B**, 38.
8. Cockayne, E. A. (1920): Proc. Roy. Soc. Med., **13**, 132.
9. Fairbank, T. (1951): *An Atlas of General Affections of the Skeleton*, p. 187. Edinburgh and London: Livingstone.

DIE MEDIESE PROFESSIE IN OËNSKOU*

J. NORTJE, M.B., Ch.B., *Voorsitter, Afdeling Noordweste van die Tak Wes-Kaapland (M.V.S.A.)*, 1959

Ek wil graag die posisie van ons professie in oënskou neem soos ek dit teen die agtergrond van die hedendaagse lewenspatroon sien—veral op ekonomiese gebied. Sowat 20 jaar gelede was toestande baie anders as vandag. Daar was minder as 'n kwart van die aantal geneeshere wat daar vandag in die Unie is. Almal was toe min of meer chronies oorwerk as gevolg van die feit dat die medisyne toe in 'n betreklike sin ondoeltreffend was in vergelyking met wat dit vandag is, en ook omdat daar so 'n groot skaarste aan aflosdokters was. Aan die anderkant is daar altyd nog 'n sterk professionele etiese kode gehandhaaf, sodat daar oor die algemeen opregte samewerking tussen dokters was. Die dokter was feitlik onaantasbaar en hy is as 'n wonderlike wese beskou—'n ideaal wat hy dan ook daadwerklik nagestreef het. Die gesinsdokter was die enigste dokter aan wie die huisgesin geglo het. Ook het dokters geweldig baie gratis dienste gelewer uit die goedheid van hulle harte sonder dat hulle ooit gevoel het dat hulle daartoe verplig was deur een of ander motief behalwe naastediens.

Gedurende die veertiger jare is die vry hospitaaldiens ingestel sonder behoorlike oorlegpleging met ons professie. Alhoewel die diens vir die publiek vry was, was dit egter om verstaanbare redes nie meer so vrywilliglik gelewer nie. Welgestelde mense kon, en het ook, gratis behandeling ontvang ten koste van die dokters. Meer inwonende dokters moes gevind word vir die hospitale en dit is toe dat die wet gemaak is dat 'n jong dokter eers minstens 1 jaar lank hospitaaldiens moet doen voordat hy mag praktiseer. Nie een van ons sal ontken dat hierdie ondervinding goed is nie, maar die salaris waarvoor die inwonende dokters, veral in die begin, moes werk, was baie onbevredigend. Die professie was magteloos om iets daaraan te doen. Toe vry hospitaaldiens in die Vrystaat ingevoer is en daar nie genoeg dokters bereid was om daarby aan te sluit nie, is die dreigement gebruik dat dokters ingevoer sou word om die betrekkings te vul.

Algaande het die ou soort gesinsdokter verdwyn en vandag gee nagenoeg 80% van die publiek nie juis veel om watter dokter hulle behandel nie. Hiermee saam het baie van die eertydse professionele etiket verdwyn of so rekbaar geword dat dit soms onerkenbaar is.

Teenoor die ongeveer 2,000 dokters van 20 jaar gelede is hier vandag amper 9,000 in die Unie. Daar is dus nou 4-maal meer dokters terwyl die bevolking nie eers verdubbel het nie. Vanaf 1954 tot 1958 het hier 1,781 geregistreer, terwyl slegs 762 uit die praktyk getree het; daar was dus 'n vermeerdering van 1,019 in 5 jaar. In 1958 is aan die verskillende Universiteite saam 1,353 mediese studente geregistreer, terwyl die getal in 1954 slegs 1,158 was; daar was dus 195 meer in 1958. Die vraag is hoe en waar almal 'n goeie bestaan sal kan maak. Die groot faktor wat ons deeglik in gedagte moet hou is dat baie Naturelle na die Naturellegebiede moet terugkeer en 'n mens sal verbaas wees om te sien hoeveel van ons inkomste ons hierdeur sal verloor.

Ons moet dus die feit in oënskou neem dat die versadigingspunt van aanvraag en aanbod in ons professie haas bereik is, indien dit nie reeds klaar so is nie. Ons moet onsself ook afvra of ons professie nie later tot 'n soort laerangse sukkelbestaan sal ontwikkel nie. Ons, met wie dit vandag goed gaan, mag nie die oë sluit en reken dat die toekoms ons nie aangaan nie, want ons is gebind aan mekaar deur professionele broederskap en ons mag ons plig teenoor ons kollegas nie versaak nie.

Ons staan alreeds sterk verenig in ons Mediese Vereniging, maar die vraag is hoeveel *mag* ons in werklikheid het. As lede van die Mediese Vereniging moet ons met die hoed in die hand ons versoeke rig om ons regte. Ons het geen wetlike mag agter ons nie. Hoeveel van ons kollegas behoort selfs nie eers aan ons Vereniging nie of betaal nie gereeld hulle ledegelde nie? Kan ons 'n krisis die hoof bied? Dit is goed om te onthou wat in Engeland gebeur het toe 95% van die dokters teen hulle sin eensklaps staatsamptenare geword het. Wat kan dit verhoed dat dit nie by ons gebeur nie?

As individue is ons onbeskermd. Dit is met teleurstelling dat ons in die dagpers soms lees dat 'n kollega weens die een of ander oortreding deur ons geagte Mediese Raad in die openbaar verhoor word. Dit werp 'n swak lig op ons professie. As 'n kollega oortree behoort hy agter geslote deure verhoor te word. Ons word nie altyd behoorlik geraadpleeg oor vraagstukke van volksgesondheid nie. Ons weet wat die beste is en moet per slot van rekening die werk doen, daarom dring ons aan op meer seggenskap en op salarisse wat by die status van ons professie pas.

As doktersgelde vergelyk word met gelde wat aan advokate en prokureurs betaal word—twee groepe van professionele mense van wie die kursusse korter is as die van dokters—dan wonder 'n mens waarom daar so 'n groot verskil is. Die werk van dokters is die gevaarlikste van alle professies of beroepe en is so veeleisend dat die lewensduur van dokters oor die algemeen betreklik kort is. Die gemiddelde winste uit ons werk regverdig nie die risiko nie. Ons het aan die toestand gewoond geword en merk dit dus nie altyd nie; maar die tyd het aangebreek dat ons beter na onsself en na ons eie sake moet kyk.

Ons het met verbasing verneem hoe die aptekers probeer het om die fundamentele reg van die dokter om self sy medisyne aan te maak te ontneem. Ek glo nie dat ons, wat hierdie vraagstuk betref, al die einde daarvan gehoor het nie; as ons nie oppas nie kan ons hierdie reg ook nog verloor.

Wat kan ons doen? Soos ek die toestand sien moet ons onsself verenig in 'n mediese professionele verbond, of wat dit ook al genoem mag word, met wetlike mag agter ons om ons besluite bindend te maak sodat ons in staat kan wees om 'n verenigde front te vorm teen alles wat ons professie in die gevaar stel. Ons moet die beginsel van ‚geslote geledere' toepas sodat slegs lede van ons verbond aanstellings kan kry. Ons moet alle hoërskoolleerlinge voorlig oor die vooruitsigte in ons professie sodat voornemende dokters nie onder enige waan verkeer wanneer hulle besluit om 'n mediese kursus te volg nie. Ook moet ons ons kollegas beskerm. Dit is baie menslik om iets vir jou eie voordeel te doen wat nie billik en professioneel korrek is teenoor kollegas nie, en hierteen moet ons waak.

As dokters het ons 'n hoë status in die samelewing—laat ons dit beskerm terwyl daar nog tyd is.

* Voorsittersrede, Strandfontein, K.P., 28 November 1959.

AMPTELIKE AANKONDIGING : OFFICIAL ANNOUNCEMENT

WYSIGING VAN VERORDENING 6 (c)

Dit word hiermee vir algemene inligting bekend gemaak dat die Federale Raad van die Mediese Vereniging van Suid-Afrika op sy sitting wat in Oos-Londen gehou is op 24 September 1959 besluit het om Verordening 6 (*c*) te wysig om soos volg te lees:

AMENDMENT OF BY-LAW 6 (c)

It is hereby notified for general information that the Federal Council of the Medical Association of South Africa at its meeting in East London on 24 September 1959 resolved that By-law 6 (*c*) be amended to read as follows:

‚6 (*c*) Lede wat die Vereniging sonder onderbreking ten minste 40 jaar lank gedien het, sal Lewenslange Lede word. Die betrokke Tak moet aansoek doen om die nodige informasie by die Hoofkantoor van die Vereniging.'

Op Las van die Raad

A. H. Tonkin
Sekretaris

Mediese Huis
Waalstraat 35
Kaapstad
21 Januarie 1960

'6 (*c*) Members who have served the Association continuously for at least 40 years shall become Life Members. The Branch concerned shall supply the necessary information to the Head Office of the Association.'

By Order of the Council

A. H. Tonkin
Secretary

Medical House
35 Wale Street
Cape Town
21 January 1960

VERENIGINGSNUUS : ASSOCIATION NEWS

CAPE WESTERN BRANCH FUND-RAISING PROJECT: GRAND PREMIÈRE

The Cape Western Branch of the Medical Association of South Africa has organized a Grand Première at the Odeon Theatre, Sea Point, in order to raise funds for the Benevolent Fund of the Medical Association. The date set for this function is Monday 29 February, and it will be held under the distinguished patronage of the Administrator of the Cape, Dr. the Hon. and Mrs. du Plessis. The film is an exciting Russian production entitled *Quiet Flows the Don.*

From 6.15 to 8.15 p.m. there will be an open-air supper party in the Sundowner Garden of the Marine Hotel, which lies immediately behind the Odeon Theatre. Attendance at this party, which is included in the ticket, will ensure convenient parking close to the theatre. If it rains on that day, the Marine Hotel will honour this part of the tickets at any time during the 10 following days. The Ladies Committee is arranging a reception for all ticket holders in the foyer of the theatre. There will also be gifts, with a Russian flavour, for lucky seat holders.

Vouchers for this evening will cost £1 1s. 0d. and may be exchanged for tickets at the mezzanine floor of the O.K. Bazaars, Cape Town, or from the Odeon Theatre booking office. Those members with preferential booking may obtain their tickets from the Cape Western Branch office. Vouchers are obtainable from the Branch office as well as from members of the Ladies Committee.

DIE LIEFDADIGHEIDSFONDS : THE BENEVOLENT FUND

Met dank word die volgende skenkings gedurende die maande September, Oktober, November en Desember 1959 erken:
The following donations during September, October, November and December 1959 are gratefully acknowledged:

Geloftekaarte ter Nagedagtenis aan: Votive Cards in Memory of:

Mrs. Bailey (Snr.) *by* Dr. H. O. Hofmeyr and family and Dr. and Mrs. R. Heald; Mrs. Mary May *by* Dr. J. Black; Mr. H. G. Willmot *by* Dr. and Mrs. W. Gilbert; Dr. M. Hoffman *by* Dr. H. W. Needham; Dr. J. P. W. Viljoen *deur* dr. en mev. J. J. Van Zyl; Mrs. C. McLean *by* Dr. L. E. Lane, Dr. and Mrs. F. E. Wynne, Dr. and Mrs. C. C. Grünberger, Drs. Tarlie, Braude and Goldblatt, Cape Midlands Branch (M.A.S.A.), Dr. and Mrs. A. S. Weir, Dr. and Mrs. J. A. Negpen, Dr. and Mrs. P. Wentzel, Medical Committee Provincial Hospital, Port Elizabeth, Dr. F. B. Kruger, and Dr. A. S. F. Murray; Mrs. E. M. McDavid *by* Dr. and Mrs. W. A. Dodds; Mrs. K. C. Lomberg *by* Prof. J. F. Brock; Dr. G. W. Doran *by* Commerce Department and Commerce Staff East London Technical College, Mr. and Mrs. W. O. S. Beauchamp, Messrs. Cooper and Cannon, Mr. G. V. Hartley, Drs. B. Navid, J. K. McCabe and H. Bloch, Mr. B. Thorne, Mr. and Mrs. C. H. Rose, Mr. C. C. Lockett, Mr. and Mrs. W. K. Pringle, Mrs. G. Ross Blaine, Mr. J. G. and Miss J. Thomas, Maureen and Charles Snyman, Mr. and Mrs. D. J. Pienaar, and Mr. A. W. Meacham; Dr. W. J. O'Brien *by* Dr. and Mrs. A. H. Baxter, Natal Inland Branch (M.A.S.A.), Dr. D. G. Cowie; Mrs. Z. A. de Beer *by* Dr. V. Brink; Dr. D. Hugo *by* Dr. L. S. Robertson.

Totaal ontvang van Geloftekaarte: £55 12s. 0d.
Total received from Votive Cards:

Dienste Gelewer aan: Services Rendered to:

Sadie De Vos *deur* Dr. P. D. Nel.
Mr. H. W. Nash *by* Dr. W. G. Schultze.
Dr. A. A. M. Coutts *by* Mr. A. Beiles and Dr. H. Curwen.
Dr. S. Rachman *by* Dr. V. Solomon.
Rev. R. M. Parker *by* Dr. L. E. Lane.
Dr. A. J. Orenstein *by* Dr. L. Lowenthal and Dr. L. Staz.
Mrs. S. Kaufman *by* Mr. L. McGregor, Dr. S. Hoffman, Prof. Murray and Dr. N. C. Leiman.
Dr. J. Walker *by* Drs. K. Pein and J. P. Grieve.
Dr. J. N. Sher *by* Drs. V. Solomon and S. Solomon.
Dr. H. E. Clifford *by* Drs. F. C. Friedlander, A. Clarke and R. J. McMahon.
Christopher Anderson *by* Dr. W. E. Owens.
Dr. J. P. H. Anderson *by* Drs. J. M. Hoffman, F. W. Morrison and B. G. Francis.
Dr. J. P. M. Retief *by* Mr. Hamilton Bell and Drs. E. Van Hoogstraten, P. S. Jenkin, I. Van Selm, L. v. d. Spuy, C. R. J. Muller and P. Maytham.
Mrs. A. J. v. d. Spuy *by* Dr. C. M. Ross.
Mrs. W. A. Tomlinson *by* Drs. P. Boshoff, D. Haynes, E. Samuels and partners.

Totaal Ontvang vir Dienste Gelewer:
Total Received from Services Rendered: £131 18s. 0d.

Skenkings: Donations:	£	s.	d.
Drs. D. S. Palmer, J. H. Boshoff, H. Euston-Brown E. Meyer, S. T. Adendorff, S. H. Daneel, A. L. v. d. Merwe	3	13	0
Medical Wives Association, Port Elizabeth, proceeds of Dinner-Dance	151	11	0
Dr. A. G. ffolliot	3	3	0
Drs. M. A. Robertson, T. McChesney and C. Kroon	6	6	0
Merry-go-round function per Mrs. W. Girdwood (Dr. D. J. Clark and Dr. A. I. Friedman)	2	2	0
Dr. W. P. Prinsloo	1	1	0
Dr. S. S. Per	1	1	0
Dr. E. O. Church	1	1	0
Dr. G. C. F. V. Helsdingen	1	1	0
Drs. Jankowitz, Blake and Wannenburg	3	3	0
Drs. Meyer, Parker and Smith	3	3	0
Drs. Warren and Zeiss	2	2	0
Potchefstroom Division (M.A.S.A.)	22	12	0
Drs. Goldberg and Kahn; Turton, Vosloo, Nienaber and Esterhuizen	6	6	0
Jones Phillipson Golf Tournament per Dr. J. V. Milner	9	10	0
Dr. J. P. Theron	3	3	0
Dr. D. M. King	4	0	0
Afdeling Oos-Transvaal (M.V.S.A.)	5	0	0
Dr. Alan Sichel Golf Competition per Dr. J. Roux	11	0	0
Campbell Cup Golf Competition per Dr. Gelb	14	15	0
Southern Transvaal Branch (M.A.S.A.) Ladies Committee per Mrs. W. Girdwood	40	2	6
Cape Western Branch (M.A.S.A.), Collection box	7	11	2
Totale Skenkings: Total Donations	£303	6	8
Groot Totaal: Grand Total	£490	16s.	8d.

IN DIE VERBYGAAN : PASSING EVENTS

Lede word daaraan herinner dat hulle die Sekretaris van die Mediese Vereniging van Suid-Afrika, Posbus 643, Kaapstad, sowel as die Registrateur van die Suid-Afrikaanse Geneeskundige en Tandheelkundige Raad, Posbus 205, Pretoria, moet verwittig van enige adresverandering. Versuim hiervan beteken dat die *Tydskrif* nie afgelewer kan word nie. Dit het betrekking op lede wat oorsee gaan sowel as dié wat binne die Unie van adres verander.

Research Forum, University of Cape Town. A meeting of Research Forum will be held on Wednesday 17 February in the Bennie de Wet Lecture Theatre, A-floor, Groote Schuur Hospital, Observatory, Cape, at 12 noon. Dr. G. G. Harrison will speak on 'Temperature changes in children during general anaesthesia'. All who are interested are invited to attend this meeting.

IN MEMORIAM

DANIEL HUGO, M.B., B.CH. (DUBL.), F.R.C.S. (EDIN.), D.O.M.S. (R.C.P. & S. ENG.)

Dr. C. C. Freed, of Johannesburg, writes:

Dr. Daniel Hugo died on Thursday, 26 November 1959 at the age of 62. It was my great privilege to have known him intimately for the past 17 years.

After a brilliant career at the University of Cape Town in which he completed his pre-medical studies, he continued his medical course at Trinity College, Dublin, where he qualified M.B., B.Ch. On his return to South Africa, he practised at Porterville, Cape Province, for many years. He then returned to the United Kingdom, obtaining the F.R.C.S. (Edin.) and the D.O.M.S. of the Royal College of Physicians and Surgeons, England. He returned to specialist practice in Johannesburg, and after some years of a highly successful ophthalmic practice, he was advised, because of the first manifestations of vascular disease, to confine himself to a less arduous branch of medicine. It was at this stage that he became the consulting ophthalmologist to the Chamber of Mines Hospital, Johannesburg.

Dr. Hugo

To evoke a living, human picture is difficult even for a word conjurer, and quite beyond my power, so probably only those who knew Daniel Hugo will be able to convert these words into a living memory, for his qualities were spiritual and hidden, and in consequence easier to feel than to describe . He was undoubtedly a great man, and no-one could know him for long without being influenced for the better. He was essentially honest in his whole approach to medicine, and could not tolerate anything approaching insincerity or hypocrisy. He had an encyclopaedic knowledge of the literature and practice of ophthalmic surgery, and his keenly critical brain enabled him to sift the wheat from the chaff very effectively. This was especially apparent when he dealt with medico-legal problems, not only in his own field of ophthalmology but also in the wider field of trauma in its relation to disease.

Among his most endearing qualities were his sense of humour, his love of life and his affection for his fellow men. Yet, he was surprisingly humble, and in consequence his remarkable ability was evident to only a comparatively limited circle. He was really a very sentimental man, but he would hide his feelings at all costs. He hated to be complimented or thanked. One grew gradually to apprehend the warmth of his personality, so that his friendship, when achieved, was deeper in consequence.

We salute his passing too soon for any of us to bear without great sorrow. To Mrs. Cynthia Hugo, whose house and hospitality were always open to us we extend our deepest sympathy.

Dr. C. F. Krige, of Johannesburg, writes:

Niel Hugo is dead. A great and good man, a wonderful friend and a magnificent doctor has departed to a welcome and well-earned rest. It is wellnigh impossible to avoid being sentimental in writing about this paragon amongst his contemporaries.

In the days of his youth he was a good student and an outstanding sportsman. He was then a leader among his fellow-students, and knew how to enjoy life. I first met him in Vienna in 1935, where he made a great impression on me, but it was 5 years later in Johannesburg where I got to know him well, and was privileged to be his friend.

Niel's personality was moulded by the happy combination of educational achievements and spiritual qualities with which he was more richly blessed than most men. With knowledge there was skill and ability. In character there was strength and loyalty, enhanced by a natural friendliness and kindness which never left him. This man had no enemies. Herein lies the secret of his success as a man and as a doctor. In the large institution which was the sphere of his activities for 16 years or more, he was revered and loved by everyone. He was friend, confidant and adviser to many a troubled soul. This he remained till the end.

Though the last years of his life were confused and complicated by the worry and frustration of chronic ill-health, he never complained but carried on in his inimitable cheerful manner, forgetting his own troubles for those of his patients. It is typical of the man that he still worked on the very day on which he took ill and died in agony.

With the passing of Niel Hugo South Africa has lost a noble and illustrious son, and our profession a truly great and good physician. He will be difficult to replace.

To all his family, our deepest sympathy. They will miss him sadly.

Ek sou graag alles in Afrikaans wou geskryf het, maar voel dat ek nie die taal magtig genoeg is om paslike hulde te bring aan Niel Hugo nie—die liewe goeie man wat so baie van ons in die lewe geken het. Ons innigste simpatie met sy bedroefde familie.

NUWE PREPARATE EN TOESTELLE: NEW PREPARATIONS AND APPLIANCES

X-RAY SCREEN-IMAGE CINE EQUIPMENT

VEB Carl Zeiss, Jena, introduce new X-ray screen-image cine equipment and supply the following information:

Medical X-ray diagnosis turns increasingly to methods of examination which, in addition to providing a radiograph of an organ under investigation, will also make it possible to draw certain information and conclusions regarding its function. For this functional radiological diagnosis apparatus is required with which serial photographs can be taken and which can be adapted according to the needs of the case to be diagnosed. The seria. photographs can be taken either according to the direct or indirect method (screen-image photographs). The indirect method of photography has the advantage that the cost of film for serial pictures remains low. The evaluation of the film strips is very easy and the frame frequencies can be obtained with moderate technical effort.

The Zeiss-Jena X-ray screen-image cine equipment consists of a transportable base on to which the angular tube and the cine

camera are mounted. By means of two set-screws the entire apparatus can be fixed to the floor during photography. In the angular tube there is a mirror which deflects the rays arriving from the fluorescent screen through 90° to the direction of the cine camera. A Neossal fluorescent screen with an antidiffusing diaphragm is used. On each side of the angular tube is a viewing window for checking the radioscopic image. Whilst observing, the covering flap is opened by turning a lever; when not in use, this closes the viewing window. For better adaption, cloth caps are provided which the observer can pull over his head. The cine camera is equipped with an X-ray lens system F/0.85 f= 120 mm. and reproduces a fluorescent screen area of 350 mm. × 350 mm. on the film at a size 56 mm. × 56 mm. (medium format). The reproduction scale $\beta = 1/6{\cdot}4$, 70 mm. wide and perforated X-ray film for screen photography is used. The built-in shutter diaphragm has a bright-field section of 240°. The stepping mechanism for the film transport has a switching ratio S = 1 : 4. To utilize fully the power of the lens system, it is essential for the film to lie continually in the same position in the one plane. Therefore, the lens has on its image side a plane-lens surface against which the film is pressed during exposure by means of a pressure plate. During film transport, however, this pressure plate as well as the film strip is lifted away from this lens surface to prevent it from being damaged or scratched.

The film cassettes can take rolls of film up to 30 metres in length. If a series of photographs has been made, the film strip can be cut with built-in film scissors and removed from the camera. The metre indicator shows the length of available film (in metres) which is still in the cassette.

To keep the radiation intensity on the patient to a minimum, it is necessary to switch off the X-ray tube during film transport. For this purpose, an electrical-impulse generator is incorporated with the camera which takes care of the control of the X-ray tube by means of control equipment supplied by specialized manufacturers of X-ray equipment.

During filming, an electrocardiogram can be made simultaneously. Each 5th picture on the film is marked on the edge with a light spot. On the electrocardiogram strip the same marking is made. Thus the film strip can be compared with the notes of the ECG, and evaluated accordingly.

For examination purposes it is recommended that the catheter be used on a special movable table, employing separate X-ray equipment for illumination and observation. After catheterization the table is moved above the cine equipment from which the wooden slab has been removed. The patient may, however, also be laid directly on the wooden slab, if a suitable table can be pushed to the cine equipment. It is advisable to lay the patient on to the table or the wooden slab so that the axis of the body is parallel to the longitudinal axis of the cine equipment.

For the evaluation of the screen image films the following instruments have been designed: evaluation of the individual pictures can be carried out with an X-ray film viewer with a magnifying lens of 1·5 magnification. If the film is to be projected as a moving picture, the X-ray Film Copying Equipment '70/35' can be used to print it on standard 35 mm. film. The copy thus obtained can then be shown with any standard film projector.

The South African representatives of VEB Carl Zeiss Jena are Messrs. Optolabor (Pty.) Ltd., P. O. Box 1820, Johannesburg.

ELESTOL

FBA Pharmaceuticals (S.A.) (Pty.) Ltd announce the introduction of a new antirheumatic and analgesic preparation Elestol, and supply the following information:

Elestol is a combination product, each tablet contains 40 mg. of Resochin (chloroquine diphosphate), 0·75 mg. of prednisone and 200 mg. of acetylsalicylic acid.

Numerous investigators have confirmed the value of chloroquine diphosphate for the treatment of rheumatoid arthritis. Its mode of action differs from all other drugs in that the effect is slow in beginning, but after the initial latent period, a marked improvement becomes manifest.

During this latent period it is often necessary to give additional analgesics and, especially, corticosteroids. The combined therapy ensures that lower corticosteroid dosages are required and that the corticosteroid effect is stabilized by the chloroquine.

Similarly, with the simultaneous administration of corticosteroid and acetylsalicylic acid, the dosage of both preparations can be reduced and the same or better therapeutic effects obtained.

With Elestol therapy, pain is usually considerably alleviated after only a few days. Subjective symptoms improve fairly rapidly even in severe cases of chronic rheumatoid arthritis. Because of the antiphlogistic properties of Elestol, the regression of acute inflammatory swellings of the peri-articular tissues can be readily observed and is associated with improvement of joint mobility.

Elestol is indicated for chronic rheumatoid arthritis, subsiding acute articular rheumatism and in rheumatic spondylitis. In osteo-arthritis associated with inflammatory symptoms Elestol may be given in addition to other treatments. It is also suitable for muscular rheumatism, humero-scapular periarthritis, and all painful inflammatory conditions of the locomotor system.

The dosage is 2 tablets 3 times a day until the condition is relieved. The tablets are swallowed whole, with some liquid, preferably after meals.

Elestol is available in boxes of 30 and 150 tablets.

BRIEWERUBRIEK : CORRESPONDENCE

ONGETROUDE VERWAGTENDE MEISIES

Aan die Redakteur: Ek sou graag deur middel van u *Tydskrif* 'n versoek rig aan medici om in gevalle waar hulle ongetroude verwagtende meisies na inrigtings verwys, die kerkverband van die meisies in ag te neem.

Die volgende is 'n lys van sodanige inrigtings in ons land:

Magdalenahuis, Claremont, Kaap.	N.G. Kerk.
Armstrong Berning-tehuis, Pretoria.	Protestants.
Irene Homes, Irene.	Anglikaans.
Salvation Army Women's Home, Johannesburg.	
Uitkoms-tehuis, Johannesburg.	N.G. Kerk.
Mary Rolt Hostel, Kaapstad.	Interkerklik.
Cotlands Babies Sanctuary, Johannesburg.	Interkerklik.
Reddingshuis van die O.V.S., Bloemfontein.	Protestants.
St. Anne's Home, Kaapstad.	Anglikaans.
Die Nannie-tehuis, Kaapstad.	N.G. Sendingkerk.

Die laasgenoemde twee inrigtings is vir nie-Blankes, en die ander almal vir Blankes.

P. O. le Roux
Sekretaris

Magdalenahuis
Claremont, K.P.
26 Januarie 1960

SOUTH AFRICAN PRACTITIONER

To the Editor: I think you will agree that your correspondent, who styles himself *Practitioner*[1] in your issue of 2 January 1960, is quite unjustified in his attack upon the *South African Practitioner* and its adoption of an ordinary, standard and recognized method of obtaining subscribers.

Your correspondent has unfairly divorced the seventh paragraph of our circular letter of 15 November from its concluding paragraph.

No attempt has ever been made to secure unwilling subscribers to the *South African Practitioner* and, indeed, to date there has been an 80% affirmative response to the circular letter, proving that the value of the publication is well recognized by the medical profession.

M. Stern
Director

South African Practitioner
P.O. Box 2395, Johannesburg
19 January 1960

1. Correspondence (1960): S. Afr. Med. J., 34, 20.

HOË TEMPERATUUR

To the Editor: In the *Journal* of 2 January Dr. Coetzee[1] of Dealesville writes about the recovery of a Native baby in whom he had recorded a rectal temperature of 109·8°F and he enquires whether such cases have been seen by others in practice.

In 1954 at the 39th South African Medical Congress in Port Elizabeth I read a paper on hyperpyrexia in infants and children which was based on the study of 153 cases where a temperature of 105·8°F or higher had been recorded. The highest temperature in my series was 110°F (rectal). This case and many others with temperatures above 107°F recovered completely (including a Native baby of 3 days with a temperature of 109·4°, where the cause also was simple overheating).

I found that the incidence in my practice was 1 in 600. The only other study of hyperpyrexia that I was able to find in the English literature was by Akerren,[2] who analysed 232 cases.

Lorn Shore

506 Medical Centre
Cape Town
18 January 1960

1. Correspondence (1959): S. Afr. Med. J., 34, 20.
2. Akerren, Y. (1943): Acta paediat. (Uppsala), 13, 449.

STERILISASIE VIR SEKSUELE OORTREDINGS

Aan die Redakteur: Vir dr. v. d. Westhuizen[1,2] is die behandeling van seksuele oortreders (‚psigopate') die eenvoudigheid self: kastreer hulle. Dit laat my baie dink aan die koningin in *Alice in Wonderland*. Vir alles het sy maar een antwoord: ‚Sny hul koppe af!'

Die verminking van die menslike liggaam is glad nie 'n nuwe verskynsel nie. In Sjina (voet-verbinding), in Basoetoland (skending van die uiterlike geslagsdele van jong meisies), in Turkye (die kastrasie van manlike harem-opsigters), ens., is dit al honderde jare lank as blote bygeloof toegepas. Om nou hierdie soorte gedoentes by ons Westerse beskawing aan te pas en ons land met eunuge te oorstroom, wat ‚gou algemeen rugbaar' sal word, sal elke regdenkende persoon met afsku vervul. Wat gaan die buiteland dan van ons dink?

Hoe kan 'n ‚psigopaat' wat gekastreer is sy selfrespek behou en 'n nuttige burger bly? My kollega erken dat die doodstraf maar min moordenaars afgeskrik het. Sal die kastrasie van seksuele misdadigers (‚psigopate') dan meer suksesvol wees? Dit sal hul net meer verbitter. By die ‚psigopaat' lê die fout in sy kop, nie in sy geslagsdele nie. Hy is 'n verskynsel van sy omgewing. Op die een of ander tyd in sy lewe het sy geestelike ontwikkeling skeef geloop sodat oordrewe sadistiese gevoelens teenoor die een of ander verskynsel in sy lewe die oorhand gekry het.

Die algemene opvatting is dat 'n mens se liggaamsbou by geboorte voldaan is; hierna word jy net groter. Maar die persoonlikheid en karaktervorming begin eers na die geboorte, en die geestelike ontwikkeling word dan grotendeels deur sy ondervinding en omgewing beïnvloed. Kan ons dan die ‚psigopaat' kwalik neem as hy nie sy abnormale gevoelens en drange kan beteuel nie en nie eens die nodige insig het om hom in alle opsigte by ons lewe en sedes aan te pas nie.

Ek stem dus baie meer saam met die redenerings van dr. Mostert[3] en dr. Bloch.[4] Wat staan ons dus te doen? In ander lande word ‚psigopate' in spesiale inrigtings opgesluit waar hulle die nodige behandeling kan geniet. Die meeste ‚psigopate' wat deur ons howe skuldig gevind word, is meer as gewillig om sodanige behandeling te ondergaan. Ons het net meer sielkundiges en psigiaters nodig, en die owerheid behoort so spoedig moontlik die nodige stappe te doen om hierdie spesialiteite aan te wakker.

Ek is dus baie verbaas om te hoor dat ‚legislation will be introduced in Parliament for sterilization or castration of sexual offenders'.[5] Is dit nodig om so kras en onverstandig op te tree? Is ons so bankrot aan gedagtes dat ons telkens ons toevlug tot wetgewing moet neem?

M. E. Dies

Kaapstad
22 Januarie 1960

1. Briewerubriek (1960): S. Afr. T. Geneesk., 34, 19.
2. *Idem* (1959): *Ibid.*, 33, 926.
3. *Idem* (1959): *Ibid.*, 33, 848.
4. *Idem* (1959): *Ibid.*, 33, 655.
5. *Idem* (1959): *Ibid.*, 33, 572.

MEDICAL AID SOCIETIES AND MEDICAL INSURANCE SCHEMES

To the Editor: It is with the greatest reluctance that I find myself compelled to comment on Dr. L. O. Vercueil's letter[1] in the *Journal* of 9 January 1960. He refers to certain unidentified colleagues 'north of the Vaal' as 'a vociferous minority—the most garrulous since the time of Hippocrates', labels them as 'agitators' and accuses them of 'disloyalty'. Dr. Vercueil should know that vilification and smearing are poor substitutes for rational arguments. Truth, logic and fair play need to be protected from whatever quarter they are assailed, so that his tirade cannot, unfortunately, be entirely ignored.

Medical Aid Societies

Dr. Vercueil's assertion that 'Medical aid societies cater for a low income group . . . most of these people would have been treated as free patients in Provincial hospitals' is patently untrue. Whatever the formula prescribed by the Association or its supposed application by the medical aid societies, the fact is that the total membership of these societies represents, typically, the white-collared class employed in the most stable, prosperous and important sectors of mining, industry and commerce in the Union of South Africa. The majority of them are the very same kind of people, earning the very same range of incomes as those whom we treat in private practice and charge customary fees for our services. With commendable foresight and thrift, the members of these medical aid societies have arranged to minimize the economic risks of illness by the payment of modest monthly contributions to their respective societies. Some groups have succeeded in getting their employers to subsidize their contributions on a £ for £ basis; some have even got their employers to pay their entire subscriptions. Members of these societies do not require, nor do they desire, any charity from the medical profession. The only concessions which they have ever sought are those which can reasonably be expected to flow from their guarantee of payment of fees in accordance with the rules of their societies.

The criticisms which have been voiced in the Federal Council and elsewhere are directed primarily against the policies of the Medical Association and not the societies. These criticisms merit careful consideration; they cannot be dismissed merely by abusing the critics. The facts are:

1. The financial responsibility of every medical aid society is limited according to the rules of that society. In some societies, the limit of liability for medical services, hospitalization and drugs is no more than £50 in any year—regardless of the cost of any major illness! Although the society generally undertakes to pay the accounts and to recover the balance from the patient, it is quite untrue to say that this is invariably so and that there are consequently 'no bad debts' or that 'medical aid societies are forced to pay 100% . . . If not recognition is withdrawn'.

2. In addition to the restrictions on the society's maximum annual liability, nearly every society pays only a proportion of each individual account out of the society's funds—sometimes as little as 40%. The rest is recoverable by the society from the patient. These limitations on the funds of the society itself mean that the main advantages accruing to a member of the society are (*a*) the loan facilities, and (*b*) the privilege of being charged for medical services at preferential rates rather than at customary rates for private patients. In much the same way, members of certain 'cooperative' societies succeed in evading the legitimate profits of the middleman by paying a small monthly contribution to the 'coop' so as to be able to 'buy retail at wholesale prices'. Since the doctor is in the position of an independent worker, not a middleman, this stratagem merely results in the exploitation of his services for lower fees without promoting, to any real extent, the principle of mutual insurance among the members of such societies.

3. The Association's schedule of preferential fees purports to represent a reduction of 33⅓% of customary private fees. This reduction is in consideration of prompt and guaranteed payment. In an analogous situation, the dental profession permits approved medical aid societies a reduction of 5%! In defence of the extraordinarily generous discount by doctors, it has been argued in the Federal Council itself, that the 'Preferential Tariff' isn't really so very preferential in respect of many of the items listed—particularly for special procedures. The moral implications of this line of argument are presumably fully appreciated by the medical

aid societies, even though they appear to have left the consciences of some of us untouched.

4. The Union-wide schedule of preferential fees for medical aid societies is a completely arbitrary and artificial invention on the part of the Medical Association. It was created to facilitate the accounting procedures of a tiny minority of medical aid societies with scattered membership throughout the Union—notably the United Banks Medical Aid Society. This schedule has since become the yardstick for private practice fees throughout the country, with consequent chaos and blatent inequity, not only for sections of the profession, but also for large sections of the public. This aspect of the problem will undoubtedly receive careful consideration by the Commission of Inquiry into the High Costs of Medical Services and Medicines recently appointed by the Minister of Health. The fact is that the fees of a general practitioner in a rural or suburban dispensing practice (such as still exist today in many inland and coastal towns and even cities), geographically situated so as to be conveniently conducted with minimal travelling from the doctor's own home, among a community which is accustomed to having most of its services —including surgery—performed by the general practioner, without making any undue demands on his time during visits or consultations, have customarily and properly always been much lower than those of his colleagues in a city such as Johannesburg ('north of the Vaal'). Here, geographical considerations and the prevailing mode of practice necessitates that the overwhelming majority of general practitioners maintain expensive consulting rooms in the centre of the city for the convenience of their widely dispersed clientele and do no dispensing and little if any surgery (seldom even the most trivial special procedure), their patients are accustomed to a pattern of examination and attention which makes more than 2 visits per hour (unremunerated for travelling), a physical impossibility.

5. Adjustment in fees for service to medical aid patients has not kept pace with monetary devaluation. The £ today has approximately ⅓ of its pre-war purchasing power. Before 1938, when the Southern Transvaal Branch of the Medical Association ran its own contract practice affairs, the fee for general practitioner's visits and consultations was 10s. 6d., travelling fees were permitted outside a radius of 3 miles and surgical fees were the same for general practitioners and specialists. Not only has a differentiation been introduced regarding surgical and other fees, but general practice fees are now £1 0s. 0d. for visits (no travelling charges permitted) and 15s. for consultations! The tardiness and reluctance with which even these inadequate adjustments have been effected by the Federal Council has led to a feeling of frustration and indignation on the part of the General Practitioners' Group of that Branch which now borders on open revolt.

6. Uniform fees for multiple visits or consultations have no parallel in private practice where repetitive visits are not only discouraged by the patients themselves, but are customarily charged for by the doctor at reduced rates. This artificial uniformity of fees is the direct result of the tariff and it is an evil thing: On the one hand, it provides an economic incentive to over-visiting and to the giving of 'courses of injections' in preference to painless and effective oral therapy. On the other hand, it places a severe penalty on diligent, detailed and time-consuming examination and treatment at the initial visit or consultation (so frequently the only attendance necessitated by the nature of the patient's complaint). The iniquitous influences of the principle of a uniform fee for multiple attendances demand the earnest attention of all who value the good name and standards of our profession.

These are some of the criticisms. Most of them, let me repeat, are directed against the Association's policies. No responsible member of our Association wishes to see medical aid societies which are genuine non-profit friendly societies abolished or discouraged, but many of us would like to see some of the absurdities of our arrangements with them set right in the interests both of the societies and ourselves and we intend to continue to 'agitate' until this is done.

Medical Services Plan

Dr. Vercueil seems to have gone out of his way to damn the Plan with faint praise. Suffice it to say that in Canada and the USA, similar medical association-sponsored plans are today the dominating influences in the control of medical practice, in the preservation of high standards of medical care and in the protection of the system of private practice from interference or exploitation by the State or by big business. Those of us who are giving much time and effort to the promotion and operation of this venture are doing so in the sincere belief that the Plan can provide a superior service to the public under conditions which are compatible with the dignity and status of our profession. The response of the public in the short period of operation of the Plan has been most encouraging. We expect and believe that our endeavours will continue to receive the loyal and unequivocal support of the most responsible members of our profession and that our Association will do nothing which may impede or vitiate the extension of the Plan on a Union-wide basis in preference to all other schemes.

Regarding the fees payable by the Plan, these have been determined by the Association itself, not by the Plan. The formula for maximum fees adopted on instruction from the Southern Transvaal Branch Council of the Association, is the medical aid schedule plus 40% (less 10% for collection). Thus, the maximum fee for a general practitioner's visit in Johannesburg is 25s. nett., not 22s. 6d. as stated by Dr. Vercueil. This formula is intended to operate until the Association is in a position to provide a valid and realistic schedule of 'customary' fees to be applied by the Plan.

Insurance Schemes

Several commercial insurance companies (notably The Crusader, SANSOM and the S.A. Mutual) have lately entered the field of medical insurance. These companies have indicated they are prepared to conduct this type of business either at a 'small profit', as in the case of The Crusader, or on a 'mutual, non-profit basis' (*sic!*) as in the case of the other two; but all have made it clear that their primary interest in this field is to expand their business in other directions—e.g. life insurance, pension funds, etc.—the contacts established by the medical insurance business serving as an introduction to other, more profitable, business. It is obvious that competition between these rival companies for the lowest possible premium rates must inevitably result in attempts to exploit the profession, the public or both. Dr. Vercueil's disclosures are the clearest indication of this inevitable trend. As he has stated, in 1958 the two 'non-profit' companies were prepared to offer guaranteed payment direct to the doctors on the basis of medical aid fees plus 25% or 33⅓% respectively. Since that time, both companies claim to have lost money—a not surprising result of their unrealistic premium rates. Now both are demanding the medical aid tariff 'plus nil'; and both have reduced benefits for future subscribers. Let it be noted that neither has sought relief by increasing premiums! One of these companies has, in addition, engaged in a deplorable propaganda campaign among its policy holders, designed to bring pressure on the doctors by their patients to accept the company's reduced cheques in full settlement of their accounts and has lately followed this up with a circular letter to all private practitioners hinting darkly at the dire consequences for the profession at large if its present demands are not acceded to. That is the position today. What will it be in a year or two if we capitulate now?

The Central Committee for Contract Practice

Dr. Vercueil demands, in effect, that we should uncritically entrust our affairs to the Central Committee for Contract Practice 'who negotiate on their behalf and who have the knowledge available and are in the best position to judge what is in the best interest of the profession and the public'. Let it be said at once that some of us believe that no committee—including the Contract Practice Committee—has any such monopoly of knowledge or wisdom. Certainly, the Contract Practice Committee carries a heavy responsibilty for recommending economic policies to be adopted by the Council. As such it must expect, and should welcome constructive criticism from every quarter and should shun any attempt to silence its critics, such as that indulged in by its erstwhile Chairman. Moreover if, as Dr. Vercueil appears to believe, the present Committee is the embodiment of his personal ideas on medical economics, it may well be entirely unrepresentative of the opinion of the profession at large.

Maurice Shapiro

5 Sasbank Building
66 Market Street
Johannesburg
19 January 1960

1. Correspondence (1960): S. Afr. Med. J., 34, 39.